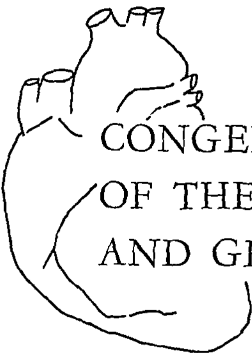
An Atlas of Congenital Anomalies of the Heart and Great Vessels



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CONGENITAL ANOMALIES OF THE HEART AND GREAT VESSELS

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Preface

THIS ATLAS was started with the concept that it would be a second edition of *Congenital Anomalies of the Heart and Great Vessels* (Dry *et al* Charles C Thomas Publisher Springfield Illinois 1948 68 pp) and in some respects it may be so considered However the expanded nature of this work has seemed to justify a designation of a different book

With exception of minor alterations all of the material presented in the earlier version is here included The basic plan of case presentation has been retained The additions represented in this atlas fall into four main categories as follows

First malformations were included which were not present in the earlier version some of these conditions being represented by cases studied in the interval between the two works Among the additional conditions included are Ebstein s malformation of the tricuspid valve cor triatriatum endocardial sclerosis partial forms of persistent common atrioventricular canal anomalous drainage of the pulmonary veins both partial and complete pulmonary arteriovenous fistula, stenosis of ostium infundibuli pulmonary stenosis with intact ventricular sep um pulmonary atresia with intact ventricular septum corrected transposition of the great vessels subaortic stenosis Marfan s syndrome and anomalies of the coronary arteries

Second there has been expansion of each of the sections dealing with the conditions presented in the earlier version In doing so use was made of the opportunity to deal in greater detail with complications of the various conditions and to show variations in cardiac or vascular structure within certain of the conditions

Third there have been included for a number of the conditions data obtained by study of patients in the Physiology Laboratory by such technics as cardiac catheterization oximetry dye dilution methods and intra arterial pressure studies Some of the cases so portrayed are purely clinical examples In o her instances there was confirmation of the clinical and physiological diagnosis either by necropsy or by operation Whenever feasible an attempt was made to include preoperative and postoperative functional studies of particular cases

Fourth the bibliography was expanded While it is not intended to be complete it gives a broad coverage of the literature on cardiovascular malformations We have arranged the bibliographic references alphabetically according to year and in each subject under the heading of the subject

The cases described in this atlas with few exceptions are from the files of several departments of the Mayo Clinic In some instances roentgenologic or other material was submitted by the patient s family physician when the patient registered at the clinic Some of these were employed if the case was selected for presentation here In other cases the family physicians were instrumental in having submitted pathologic material on patients who had been studied at the clinic during life and who had succumbed to their disease after returning home For these services we are grateful We are also indebted to those clinicians and pathologists who submitted pathologic specimens and data on patients who had at no time been seen at the clinic All of the material submitted from whatever source in addition to our own material represented a convenient and valuable collection of specimens and data from which selection was made to prepare this atlas

The individuals who submitted material are Drs F H Adams D L Alcott W S Alexander J D Barger J M Baty R C Bing E F Bland C W Borden W B Chamberlin Jr G J Cunningham J F Dammann J R Dawson L H Domeier A C Ernstene S E Gould M L Greenberg G D Griffin J C Henthorne C H Hollis Bryan Hudson L S Jolliffe E G Kidd D P Ladewig T Leary S W Lippincott T E Ludden G K Mallory Gertrude Moore J T McClellan Catherine A Neill D A Nickerson John Noble J W Old G S Owen Frederic Parker Jr M M Patton J V Pischieri W A Ricker Jr H M Rogers W O Russell Fred Sloan L A Stapley Henry Swan E T Thorsness Louise Weigenstein A H Wells and Harold Wood Whenever material from these sources was used in the atlas it has been specifically credited

Dr H Milton Rogers who was a co author of the earlier work completed his Fellowship at the Mayo Foundation prior to the preparation of this publication We are grateful to him for his contribution in the

basic work that led to the development of the previous version much of which as indicated is among the material *within this atlas*

For the preparation of this atlas the authors are indebted to many in the Mayo Clinic for help in a variety of ways all of which led finally to the completion of this publication Credit is due the other members of our respective sections as well as the thoracic surgeons the pediatricians and the roentgenologists who took part in the work up of many of the cases presented in this atlas Particular credit is due Mr Russell Drake and his Art Studio Staff All of the drawings employed in this book were prepared in his department and the majority were done by Mr Drake himself

The Photographic Department performed a service of great magnitude For this and the spirit with which it was done the authors are indebted to Mr Leonard Julin and his entire staff Much credit and thanks go to Mr Thomas Keys head of the Library and to the Library staff for assistance with matters pertaining to references To the Editorial staff go our thanks for assistance with the final stages of preparation of the manuscript and for advice during all stages of its evolution

To one who will study the pages of this atlas it will be obvious that in each of the departments with which each of us is associated there are many individuals who did the leg work the little things without which completion of this work would have been long delayed and difficult A list of all these would be impossible to make We should like however to give particular recognition to several of these people In the Section of Pathologic Anatomy Mrs Robert T Hood assisted materially in the composition of the manuscript In the same department Miss Pearl C Knutson and in the Section of Cardiology Mrs Merle Boler assisted in the assembly of the material Mr William Mayo in the Section of Pathologic Anatomy was together with members of the Photographic Department responsible for taking photographs of the gross specimens Credit for assembly of the electrocardiograms goes to the technical staff of the Electrocardiographic Laboratory The devoted efforts of the technical staff of the Cardiovascular Section of the Physiology Laboratory played a considerable role in the accumulation of data in many cases of cardiac disease some of which were used in this atlas Miss Janet Robinson prepared the illustrations of the physiologic recordings utilized herein

Models illustrated in the form of color plates were produced in the laboratories of the Mayo Foundation Museum of Hygiene and Medicine under the direction of one of the authors (Bulbulian) Some of these models are direct casts from actual specimens the others are hand modeled using the actual specimens as guides Credit for this phase of the work must be given to Mr Leonard Knudson and Miss Neta Case

Rochester Minnesota
August 1953

THE AUTHORS

Contents

An Atlas of Congenital Anomalies of the Heart and Great Vessels

Cor Biloculare

(Two chambered Heart)

COR BILOCULARE represents the most primitive type of heart encountered in man There is failure of formation of the atrial and of the ventricular septa As a rule there is a common atrioventricular valve and usually a single artery leaves the heart This artery is either a true persistent truncus arteriosus or an aorta associated with an atretic pulmonary trunk With rare exceptions patients with this condition fail to survive infancy

Cor Biloculare

(Two chambered Heart)

THIS IS THE most primitive type of heart in our series It has a single atrium a single ventricle and a common atrioventricular valve A single functioning arterial trunk the aorta leaves the common ventricle

The pulmonary artery lies behind the aorta and is atretic The circulation to the lungs is by way of a patent ductus arteriosus

The common atrium receives blood from the lungs through the pulmonary veins and peripheral blood through the inferior vena cava and a right as well as a persistent left superior vena cava No coronary sinus as such exists in this heart

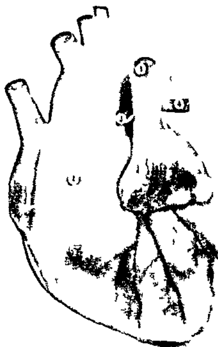


Fig 1—Left anterior view (model x1) (1) Aorta arising from common ventricle (2) Atretic pulmonary artery (3) Persistent left superior vena cava (4) Left pulmonary artery

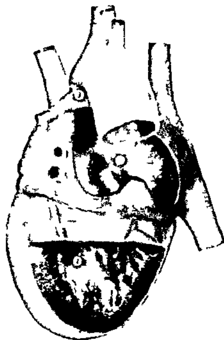


Fig 2—Left posterior view (model x1) (1) Rudimentary septum in common atrium (2) Interior of common ventricle (3) Patent ductus arteriosus

History of the Patient

FEMALE eight months old History of cyanosis since birth on feeding or crying No murmurs Hemoglobin 20 7 gm per 100 cc of blood Admitted to hospital for repair of diaphragmatic hernia Roentgenogram of thorax showed diaphragmatic hernia and prominence of right upper border of the heart Cyanosis increased postoperatively the patient died later the same day

Principal Clinical Features of This Anomaly

1 Early and progressive cyanosis
2 Occasionally no murmurs
3 Early cardiac enlargement of globular shape
4 Usually death in infancy or early childhood
5 Isolated dextrocardia may be associated



Fig 3—Specimen from which model shown in Figures 1 and 2 was prepared Interior of common ventricle and aortic orifice

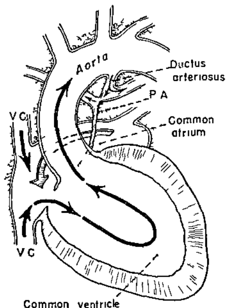


Fig 4—D ag am of intracardiac c culat on in cor b loculare with atretic pulmo ary artery and patent ductus a ter osus

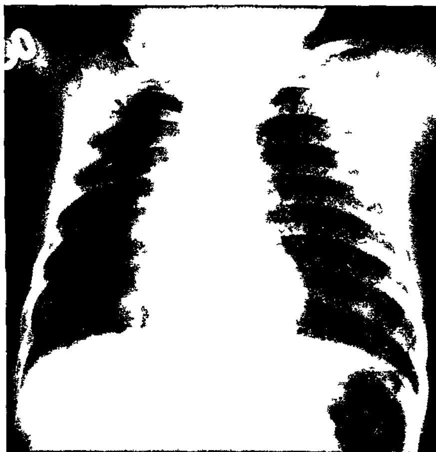


Fig 5—Tho acic roentgenogram of the p tient whose heart is llustrated in Figures 1 2 3 and 4

Cor Biloculare, Transposition of the Great Vessels, Subpulmonary Stenosis, Right Aortic Arch with Right-sided Descending Aorta and Right Ligamentum Arteriosum

THE PATIENT whose heart is illustrated on this page was a male two months old at the time of death The patient had presented a feeding problem Cyanosis was noted terminally (Specimen submitted by Dr Donald L Alcott)





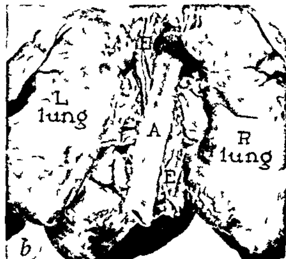


Fig 6—(above)—a The heart and lungs from the front Right aortic arch Branches mirror image of normal R C C = right common carotid artery R S A = right subclavian artery L S A = left subclavian artery L C C = left common carotid artery *b* roentgenogram of thorax

Fig 7—(left)—Interior of common ventricle Behind wide aortic origin is narrow ostium (in circle) leading to a stenotic subpulmonary tract

Fig 8— (below)—a Right anterior view of heart and great vessels The ligamentum arteriosum (Lig art) extends from right pulmonary artery (R P A) to the aorta opposite the origin of right subclavian artery (R S A) Pulmonary trunk (P T) narrow and behind and parallel to the aorta *b* Thoracic organs from behind Upper part of descending aorta to the right of esophagus Aorta crosses behind esophagus to the left in lower part of thorax A = aorta E = esophagus





Cor Triloculare Biatriatum

(Three chambered Heart)

COR TRILOCULARE BIATRIATUM is characterized by failure of formation of the ventricular septum so that the ventricular part of the heart is common to both circulations This means that arterial blood and venous blood become mixed before leaving the heart The atrial septum is formed and as a rule the tricuspid and mitral valves are also formed each communicating with the common ventricle Usually two vessels leave the heart and except in rare cases there is transposition of the great vessels In many instances there is narrowing of the outflow tract of the common ventricle below the aortic orifice occasionally there is a subpulmonic narrowing The chances of survival are greater than in cor biloculare although death during infancy is common but some patients reach adult life

Cor Triloculare Biatriatum

(Three chambered Heart)

THIS IS A three chambered heart with two atria and a single ventricle The latter represents a primitive state in which there is no ventricular septum There is transposition of the aorta and pulmonary artery as shown by the right anterior position of the aortic origin In this specimen there also is isolated dextrocardia

Fun tionally this heart varies little from a two chambered heart since there is a free mixture of venous blood and aerated blood in the common ventricle



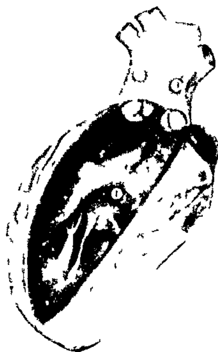


Fig 9—(left)—Anterior view (model x1) (1) Apex of common ventricle directed to the right (2) Right atrium (3) Left atrium

Fig 10—(right)—Interior of common ventr cle (model x1) (1) Common ventricle (2) Aorta aris ng anteriorly (3) Pulmonary trunk arising posteriorly

History of Patient

FEMALE 10 months old well until age of 5 months Then a cold developed with fever and respiratory diffi culty At 7 months cyanosis was noted which became more intense at 9 months Patient was admitted with pneumonia and congestive heart failure Roentgenologic study revealed isolated dextrocardia with marked cardiac enlargement Hemoglobin 17 5 gm per 100 cc of blood erythrocytes numbered 5 710 000 per cubic millimeter Cyanosis increased markedly before death

Principal Clinical Features of This Anomaly

1 Cyanosis develops early but may be mild or transient and intensified by activity The oxygen s turation of arterial hemoglobin always is sub normal 2 Systolic murmur over the entire precor dium usually is present 3 Precordial thrill may be present 4 Oxygen saturation of hemoglobin in ventricular blood exceeds that of hemoglobin in mixed venous blood (cardiac catheterization) 5 Roentgenologic aspects unusual globular configura tion (the most frequent anomaly associated with iso lated dextrocardia) 6 Electrocardiogram usually high voltage diphasic QRS complexes



Fig 11—Specimen from which models shown in Figures 9 and 10 were prepared Interior of common ventricle and origin of aorta

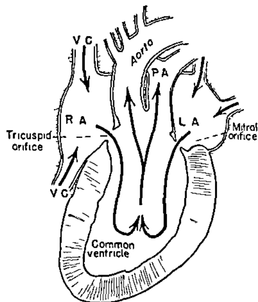


Fig 12—Diagram of ntracardiac circ lation in cor triloculare biatriatum

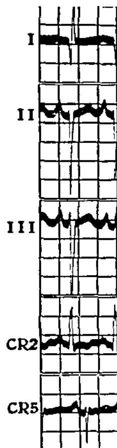




Fig 13—Electrocardiogram and thorac c roentgenogram of p t ent whose heart s illustrated F gures 9 10 11 and 12

Cor Triloculare Biatriatum with Subaortic Stenosis

The patient was a 5 month old girl who was acyanotic until shortly before death There was a rough systolic murmur loudest in the second left intercostal space



Fig 14a—The exterior of thoracic organs from the front Transposition of great vessels Aorta (A) and pulmonary trunk (P T) parallel Aorta relatively narrow pulmonary trunk wide There is also coarctation of the aorta (Coarct) and tubular hypoplasia of the aortic arch between the left subclavian artery (L S A) and the left common carotid artery (L C C) Lig art = ligamentum arteriosum



Fig 14b—The common ventricle and great vessels Probes lie in the mitral and tricuspid orifices The outflow portion of the common ventricle is separated into a narrow subaortic pocket which lies beneath the aortic valve (A V) and a wider pocket beneath the pulmonary valve (P V)



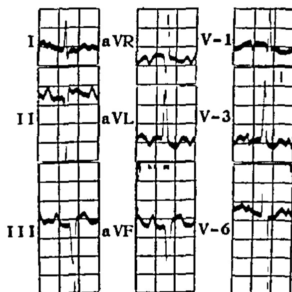


Fig 15—The thoracic roentgenogram and electrocardiogram of the patient whose heart is illustrated in Figure 14 (Roentgenogram from Rogers H M and Edwards J E *Am Heart J* 41 299 1951 with permission)

Dynamics of the Circulation in Cor Triloculare with Subaortic Stenosis

In this anomaly the common ventricle supplies the ejectile force for both systemic and pulmonary circulations (*Fig 16a*) In this way the single ventricle in postnatal life has a similar function to the right ventricle of the normal fetus (Fig 16*b*) Under each of these conditions adequate systemic blood flow depends on a relatively high resistance to blood flow in the pulmonary circulation Otherwise the lesser circulation would become flooded This high resistance is related to demonstrable structural changes in the muscular arteries of the lungs

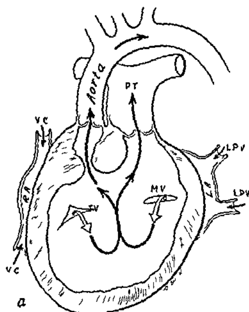


Fig 16a—The intracardiac circulation in cor triloculare biatriatum with subaortic stenosis

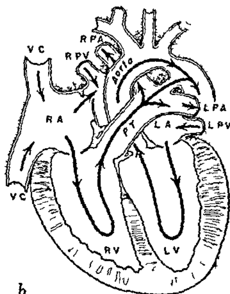


Fig 16b—The intracardiac circulation in the normal fetus (From Edwards J E and Chamberlin W B Jr *Circulation* 3 5 4 1951 with permission)



Fig 17—Photomicrographs of muscular arteries of the lungs (From sections stained with Verhoeff's elastic tissue stain counterstained with van Gieson's connective tissue stain x75) *a* A muscular artery from a normal 7 year old girl showing a thin wall and wide lumen *b* From a 6 year old boy with cor triloculare biatriatum and subaortic stenosis showing hypertrophy of the media and corresponding luminal narrowing In this patient there were no intimal changes in vessels of this size *c* From an 8 year old boy with cor triloculare biatriatum and subaortic stenosis As in the case illustrated in *b* there is medial hypertrophy but there is in addition a pronounced degree of superimposed intimal fibrosis The latter accentuates the narrowing in these vessels The great degree of stenosis caused by the fibrous tissue is fixed and irreversible (From Edwards J E and Chamberlin W B Jr *Circulation* 3 524 1951 with permission)

Atresia of the Tricuspid Orifice

(Tricuspid Atresia)

(Functional Two chambered Heart)

In this heart there is no communication between the right atrium and the right ventricle The foramen ovale is patent

The right ventricle is diminutive and communicates with the large left ventricle through a slitlike defect in the ectopic ventricular septum The pulmonary trunk is narrow the aorta wide The ductus arteriosus is closed



Fig 21—Anterior view (model x1) (1) Large left ventricle (2) Diminutive right ventricle (3) Pulmonary trunk



Fig 2—Interior viewed from right (model x1) (1) Patent foramen ovale the only outlet of right atrium (2) Large left ventricle (3) Diminutive right ventricle Probe is inserted through ventricular septal defect

History of the Patient

Female 4 months old had transient cyanosis at birth more apparent at 5 weeks of age Basal systolic murmur Readmitted at 10 weeks of age Same murmur present moderate cyanosis hemoglobin 17 7 gm per 100 cc of blood erythrocytes 5 170 000 per cubic millimeter Roentgenogram globular enlargement with absence of shadow of conus arteriosus and diminished hilar markings Electrocardiogram left axis deviation At 4 months right hemiplegia developed Patient readmitted in coma and died Thrombosis of longitudinal and transverse venous sinuses with left cerebral infarction

Principal Clinical Features of This Anomaly

1 Progressive cyanosis from birth
2 Severe spontaneous dyspneic attacks have been noted in some cases
3 Usually basal systolic murmur
4 Roentgenogram cardiac enlargement Configuration may resemble that in tetralogy of Fallot
5 Electrocardiogram left axis deviation precordial leads indicate dominant left ventricle These are important diagnostic features



Fig 23—Tricuspid atresia type I*b* Specimen from which models in Figures 1 and 22 were prepared *a* The interior of the large left ventricle This communicates freely with the aorta A narrow slitlike opening (containing probe) leads to the hypoplastic right ventricle *b* The hypoplastic right ventricle The probe lies in the tract leading from the left ventricle The narrow pulmonary trunk takes origin from the hypoplastic right ventricle (From Edwards J E *et al J T ch M thod* 28 34 1948 with permission)

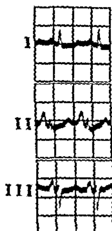




Fig 24—Electrocardiogram and thoracic roentgenogram of the patient whose heart is illustrated in Figures 1 2 and 3

Tricuspid Atresia, Anatomic Classification

THERE ARE four anatomic types of tricuspid atresia An atrial septal defect is present in all types usually this is a patent foramen ovale which is the only outlet for blood coming to the right atrium through the great systemic veins Through this opening venous blood enters the left atrium where it mixes with oxygenated blood returning through the pulmonary veins The mixture flows through an enlarged mitral valve into a large left sided ventricular chamber The flow of blood from this chamber follows one of four different routes depending on the type of tricuspid atresia (Fig 25)

Fig 25a—Tricuspid atresia with concomitant pulmonary atresia and without transposition (type I*a*) The right ventricle is excluded from the circulation and blood reaches the lungs through a patent ductus arteriosus This is an uncommon form

Fig 25b—Tricuspid atresia with subpulmonary stenosis and without transposition (type I*b*) Blood reaches the lungs through a slitlike muscular walled tract (representing the zone of subpulmonary stenosis) which connects the large left sided ventricular chamber with the small right chamber and then through the pulmonary trunk This is the commonest type of tricuspid atresia (Fig 21) Death in infancy or early childhood is usual

Fig 25c—Tricuspid atresia with subpulmonary stenosis and with transposition (type II*a*) The stenosis is either at the pulmonary valve or as is commoner just below the pulmonary valve Some blood reaches the lungs through dilated bronchial arteries This type of tricuspid atresia has the best prognosis

Fig 25d—Tricuspid atresia with transposition of the great vessels but without pulmonary or subpulmonary stenosis (type II*b*) In this form blood flows freely into the pulmonary trunk

Types I*a* and *b* and II*a* are functionally like the tetralogy of Fallot in that there is pulmonary stenosis and a venous arterial shunt Type II*b* is functionally like the Eisenmenger complex in that there is no gross barrier to pulmonary blood flow and there is a common ventricular ejectile force for the systemic and pulmonary circulations (Fig 25*b* and *d* from Edwards J E *Arch Surg* *61* 1103 1950 with permission)

Tricuspid Atresia, Type IIa

Boy 12 years old Cyanosis since birth accentuated by exercise Clubbing of digits No thrills or murmurs



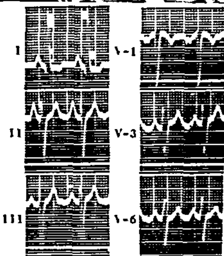


(From Edwards J E and Burchell H B *M Clin North America 33* 1177 1949 with permission Case also reported by Rogers H M *et al* *Am J Dis Child* 80 427 1950)

Fig 26—(above)—a Left sided ventricular chamber and aorta At the point of the arrow between the crista supraventricularis (C S) and the anterior leaflet of the mitral valve (M) and behind the aortic orifice is the stenotic ostium of a tract leading to the pulmonary trunk *b* Close up view of the subpulmonary ostium illustrated in *a*

Fig 27—(right)—The electrocardiogram

Fig 28—(below)—The anteroposterior and left anterior oblique roentgenograms (From Rogers H M *et al* *Am J Dis Child* 80 427 1950 with permission)







Tricuspid Atresia, Type IIb, Transposition of Great Vessels No Pulmonary Stenosis





Fig 29—Tricuspid atresia type II*b* From a boy 4 months old Specimen submitted by Dr Walter Ricker (From Edwards J E and Burchell H B *M Clin North America 33* 1177 1949 with permission) *a* Right anterior view Transposed great vessels The pulmonary trunk (P T) is wider than the aorta (A) *b* Pulmonary trunk (P T) has been opened along its right side It is in free communication with the left sided ventricular chamber There is no pulmonary or subpulmonary stenosis The probe extends from the large ventricular chamber into the space by which the two ventricular chambers communicate and disappears in the unopened aorta

Functional Studies in a Case of Tricuspid Atresia

MAN 35 years old with persistent cyanosis and electrocardiographic evidence of left ventricular hypertrophy (Data from Geraci J E *et al Proc Staff Meet Mayo Clin 23* 510 1948 with permission)

Catheterization Data

Source of sample	Oxygen content volumes per cent	Oxygen saturation per cent	Pressure mm of mercury *
Inferior vena cava	22	65 7	5 13
Pulmonary vein	3 3	98 7	6-14
Left atrium	6 2	77 5	5 13
Left atrium	26 6	9	2 14
Right atrium (middle)	19	57 0	4 18
Superior vena cava	0 0	59 8	5 15
Oximeter value for arterial oxygen saturation during catheterization		79 5	

Oxygen consumption 282 0 cc per minute surface area 1 68 square meters

* The values given represent the low and high pressures recorded

Ebstein's Malformation of the Tricuspid Valve

Ebstein s malformation of the tricuspid valve is a rare condition characterized by attachment of the septal and posterior leaflets of the tricuspid valve to the right ventricular wall at its apex The anterior leaflet is normally attached to the annulus fibrosus The abnormal valvular attachment causes the tricuspid orifice to be reduced in size The resulting small tricuspid orifice may function without associated tricuspid insufficiency Another feature of the abnormal valvular attachment is that the greater portion of the right ventricle forms a large common receiving chamber with the right atrium The only portion of the right ventricle that functions as such is the anatomic outflow portion of this chamber

An atrial septal defect is commonly associated with Ebstein s malformation and when present a venous arterial shunt may exist which may be associated with varying degrees of arterial oxygen desaturation and cyanosis

Progressive cardiac enlargement may be present A systolic and occasionally a diastolic murmur is heard over the sternum

Roentgenograms show enlargement of the right atrium and ventricle Roentgenoscopy reveals decreased amplitude of pulsations of the right side of the heart The pulmonary fields are clear and there is decreased pulmonary arterial pulsation

Angiocardiography may show a common right atrioventricular chamber

The electrocardiogram is variable and may show delayed arterio ventricular conduction right bundle branch block and huge P waves

Patients with Ebstein s malformation usually survive to adulthood The average survival period is about 25 years

Ebstein's Malformation of the Tricuspid Valve

In this heart the septal and posterior leaflets of the tricuspid valve are attached abnormally low In this way the right atrium and the sinus portion of the right ventricle form a large receiving chamber while that part of the right ventricle which functions as such is only the outflow portion of the right ventricle







Fig 30—(top left)—Exterior of heart anterior view (model x3/4) (1) Right atrium (2) Dilated inflow portion of right ventricle (3) Pulmonary trunk

Fig 31—(top right)—Right posterior view of interior of right atrium and inflow portion of right ventricle (half size model) (1) Edge of atrial septal defect (2) Anom*alously attached septal and posterior tricuspid leaflet* (3) Anterior tricuspid leaflet

Fig 32—(left)—Outflow portion of right ventricle (half size model) (4) Anterior tricuspid leaflet (5) Right auricular appendage (6) Pulmonary trunk

History of the Patient

Boy 6 years old cyanosis since birth Susceptible *to upper respiratory infections marked fatigue and* dyspnea with slight exertion Systolic thrill and murmur maximal in left fourth interspace Roentgenologic examination disclosed right ventricular enlargement and prominence of the conus arteriosus shadow but with clear lung fields Electrocardiogram revealed right axis deviation exaggerated P waves in leads II and III Angiocardiography rapid filling of left ventricle and aorta delay in filling of pulmonary artery Death during induction of anesthesia for cardiac exploration

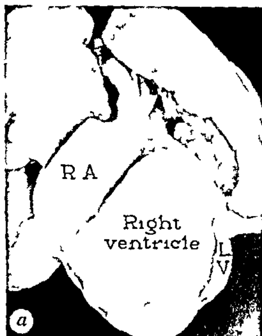




Fig 33a and b—The specimen from which models n Fig re 30 32 were prepared *a* Anterior view of unopened thoracic organs Ex ept at its apical portion the outer surface f the r ght ventricle bulges above the surface of the left ventricle (L V) Right auricular appendage (R A) dil ted *b* Interior of right side of heart Atrial septal defect Only the anterior tricuspid leaflet (A T) i attached to the ann lus fibrosus The rema nder of the tr cuspid valvular tissue (T V) is irregularly adherent to the apical portion f r ght ventricle Most of right ventricle forms a large receiving chamber with the r ght atrium

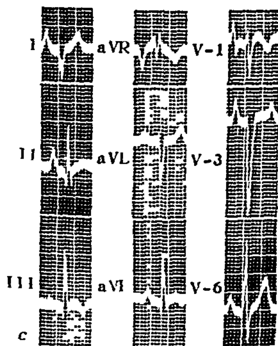




Fig 33c and d The electrocard ogram and the roentgenogram of the thorax of the patient whose heart is illustrated in Figures 30 34



Fig 34a—The outflow portion of the right ventricle from the case presented on pages 18 21 Tricuspid orifice (T O) small and guarded by the anterior tricuspid leaflet (A T) the only tricuspid leaflet that is normal This is the only portion of the heart that functions as the right ventricle P T = pulmonary trunk

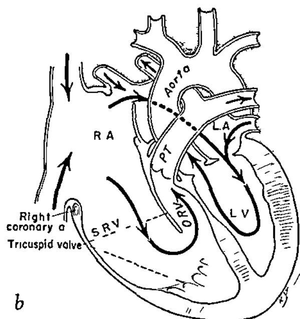


Fig 34b—Intracardiac circulation in Ebstein s malformation of the tricuspid valve associated with an atrial septal defect R A = right atrium S R V = sinus of right ventricle O R V = outflow tract of right ventricle P T = pulmonary trunk L A = left atrium L V = left ventricle





Fig 35—Angiocardiograms in the case presented on pages 18 21 *a* Taken $1\frac{1}{2}$ seconds after injection of radiopaque material *b* One half second after *a* Rapid filling of the left atrium and ventricle and the aorta (Angiocardiographic studies performed by Dr D G Pugh)

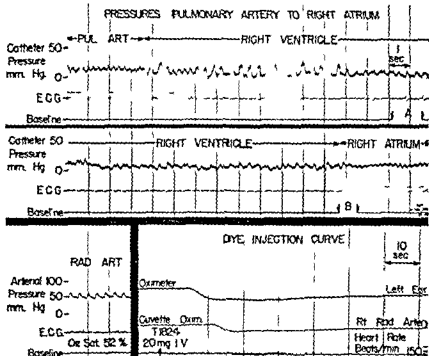


Fig 36—Functional studies in the case described on pages 18 0

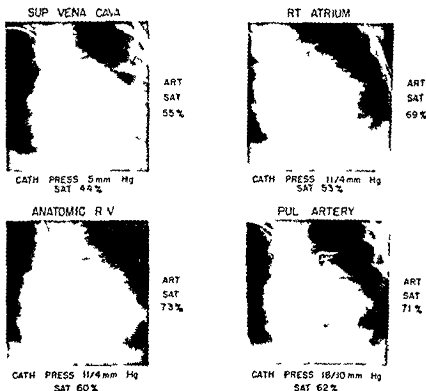


Fig 37—Roentgenograms and data obtained during cardiac catheterization in the case described on pages 18 20

Ebstein's Malformation, Abscess of Pons

MAN 30 years old Moderately good exercise tolerance Noticeable cyanosis on exertion Terminal symptoms of pontine abscess (From Barger J D *et al* *Am J Clin Path* 21 576 1951 with permission)



Fig 38a—Right atrium and ventricle Only the anterior tricuspid leaflet (A T) is normally attached Septal and posterior tricuspid leaflets (T V) attached to right ventricular wall do not function Atrial septal defect (Insert) Pons abscess No inflammatory disease of heart

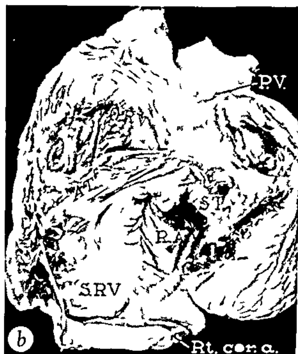


Fig 38b—Outflow of right ventricle (O R V) Anterior tricuspid leaflet (A T) normal Remainder of tricuspid valvular tissue attached to right ventricular wall P V = pulmonary valve S T = septal leaflet tricuspid S R V = posterior tricuspid leaflet attached to the sinus of the right ventricle R A = right atrium

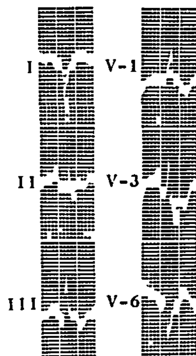


Fig 39—The electrocardiogram and the roentgenogram of the thorax

Atresia of the Mitral Orifice

(Mitral Atresia)

(Functional Two-chambered Heart)

IN ATRESIA of the mitral orifice no mitral valvular tissue is found in most instances and as the name implies there is no connection between the left atrium and the ventricular portion of the heart The route of the circulation is opposite in direction to that in tricuspid atresia Oxygenated blood from the left atrium flows usually through an opening in the atrial septum into the right atrium where it mixes with venous blood In rare instances the foramen ovale is closed and an anomalous vein extending from the left atrium to a systemic vein (levoatriocardinal vein) represents the route of exit for left atrial blood Whatever the pathway of exit for left atrial blood it is usually inadequate and in the presence of mitral atresia there exists a barrier to pulmonary venous blood flow

The mixture in the right atrium flows through a large tricuspid orifice into the ventricular portion of the heart In some cases there are two ventricles and a ventricular septal defect and in other instances there is a common ventricle Transposition of the great vessels is frequently associated Survival beyond infancy is uncommon

Atresia of the Mitral Orifice

(Mitral Atresia)

(Functional Two chambered Heart)

In this heart there is no communication between the left atrium and the left ventricle The foramen ovale is patent

The left ventricle is small and communicates with the larger right ventricle through a defect in the membranous portion of the ventricular septum

The aorta arises anteriorly and to the left and is slightly narrower than the pulmonary trunk The ductus arteriosus is closed

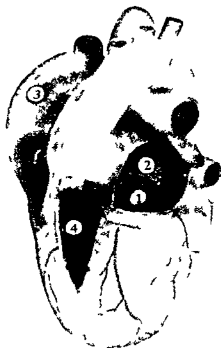


Fig 40—Left lateral view (model x1) (1) Atresia of mitral orifice (2) Patent foramen ovale (3) Large right atrium (4) Small left ventricle

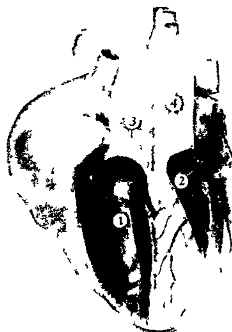


Fig 41—Anterior view (model x1) (1) Large right ventricle (2) Small left ventricle (3) Pulmonary trunk (4) Aorta

History of the Patient

Male 4 months old Loud systolic murmur at birth Slight intermittent cyanosis beginning at 2 months of age Child presented a feeding problem and there was poor development On final admission loud systolic murmur and thrill were noted over entire precordium and interscapular area Roentgenogram showed marked cardiac enlargement the electrocardiogram showed right axis deviation Severe terminal cyanosis with pneumonia

Principal Clinical Features of This Anomaly

1 Progressively severe cyanosis
2 Infant may be dyspneic and weak
3 Early development of cardiac failure
4 *A systolic murmur is usually present*
5 Roentgenologic aspects marked right ventricular enlargement and prominent shadow of pulmonary artery
6 Electrocardiogram persistence of infantile type of electrocardiogram with picture of right ventricular hypertrophy
7 Death usually occurs in early infancy



Fig 4 —Specimen from which models shown in Figures 40 and 41 were prepared Interior of left atr um Blind dimple at expected location of mitral valve

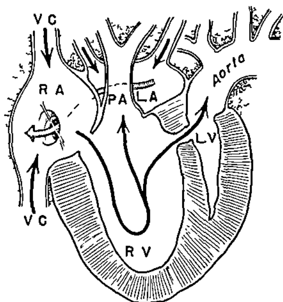


Fig 43—D agram of intracardiac circulation in atresia of mitral orifice

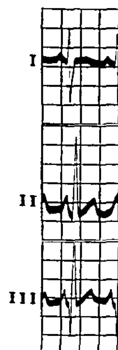




Fig 44—Elect oc diog am and thoracic roentgenogram of the pat ent wl ose heart is illustrated n Figures 40-43

Atresia of Mitral Orifice, Common Ventricle and Transposition of Great Vessels

FEMALE 6 weeks old Cyanosis since birth Systolic precordial murmur and cardiac enlargement Progressive dyspnea and terminal cardiac failure



Fig 45—Exterior of heart and great vessels Transposition of great vessels Tubular hypoplasia of aortic isthmus L S A = left subclavian artery A = ascending aorta P = pulmonary trunk L P A = left pulmonary artery R P A = right pulmonary artery Lig art = ligamentum arteriosum



Fig 46—Interior of common ventricle The outflow tract of the common ventricle divided into a narrow subaortic portion (Sub ao) and a larger subpulmonary portion (probe) Tricuspid valve (T V) opens into sinus of common ventricle Inherent in the condition there is no mitral orifice A patent foramen ovale allowed flow of blood from the left atrium into the right

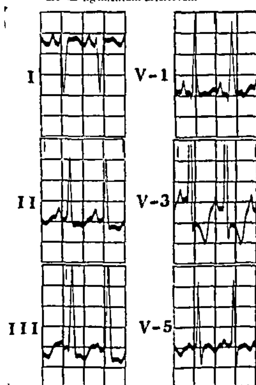




Fig 47—The electrocardiogram and the thoracic roentgenogram

Cor Triatriatum

(Triatrial Heart Congenital Stenosis of Common Pulmonary Vein)

COR TRIATRIATUM is another congenital anomaly causing a barrier to the emptying of the pulmonary veins In this condition the pulmonary veins empty into an accessory chamber lying superior to the true left atrial chamber and communicating with it by means of a narrow opening The narrow opening between the accessory chamber and the true left atrium constitutes a point of obstruction to pulmonary venous flow Functionally the condition resembles mitral stenosis

The accessory chamber seems to represent the common pulmonary vein of the embryo It has failed to become incorporated into the left atrium as it normally should

Cor Triatriatum

In this specimen the pulmonary veins empty into an accessory chamber lying above the true left atrium The accessory chamber and the left atrium communicate by means of a narrow opening

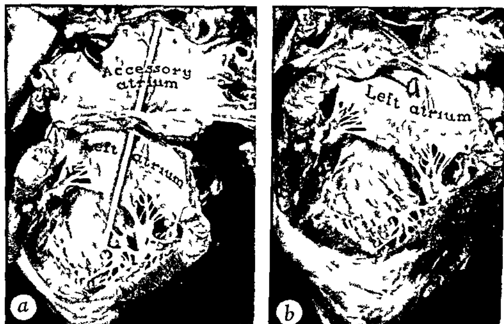


Fig 48—The left side of the heart *a* The pulmonary veins enter the accessory atrial chamber lying above the true left atrium The opening between the two chambers i narrow (see *b*) *b* The narrow opening (containing probe) between the accessory chamber and the true left atrium as viewed from below

History of the Patient

Female 6 months old Cyanosis of lips periorbital areas and nose noted at birth Irritability frequent vomiting and diarrhea and feeding difficulties Attacks of screaming rigidity pallor and lethargy two weeks prior to admission Increasing dyspnea Lips and nails good color heart beat rapid but regular no murmurs lungs clear liver enlarged to umbilicus No cyanosis Died suddenly on the morning following admission (From Edwards J F *et al* *Arch Path* *51* 446 1951 with permission)

Principal Clinical Features of This Anomaly

1 Respiratory embarrassment due to pulmonary congestion from an early age (cyanosis may appear owing to pulmonary congestion or cardiac failure)
2 Enlargement of the right ventricle and upper chamber component of the left atrium eventually right heart failure
3 Absence of murmurs or presence of diastolic apical murmur tachycardia
4 Electrocardiogram expected to show evidence of right ventricular hypertrophy

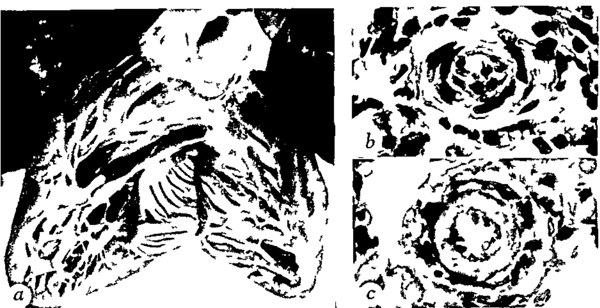


Fig 49—From the patient whose heart is illustrated in Figure 48 *a* The hypertrophied right ventricle *b* A pulmonary arteriole shows intimal cellular fibrous proliferation (H & E x 600) *c* A small pulmonary muscular artery shows medial hypertrophy and a prominent internal elastic membrane (Verhoeff's elastic tissue stain counterstained with van Gieson's connective tissue stain x 740) The vascular lesions are considered to be effects of impaired venous drainage from the lungs and resembled those seen in mitral stenosis

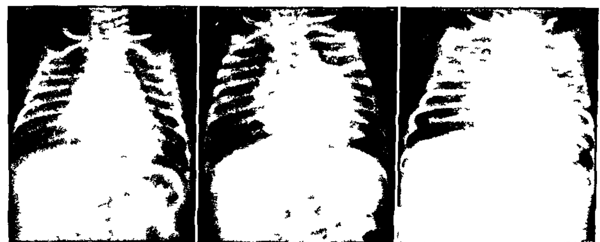


Fig 50—Roentgenograms of the thorax of the patient whose heart is illustrated in Figures 48 and 49 (Reproduced by courtesy of Dr W N Doss)

Cor Triatriatum, Probable Developmental Basis

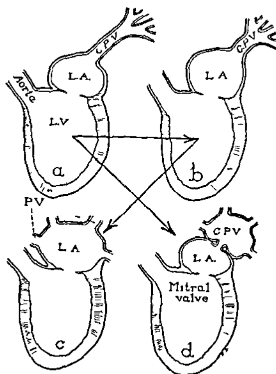


Fig 51—In *a* *b* and *c* are demonstrated the stages by which the common pulmonary vein of the embryo is normally incorporated into the dorsal wall of the left atrium In *d* there is shown an arrest of this process leaving the common pulmonary vein distinct from the true left atrium Because of the stenosis at the junction of the two chambers the common pulmonary vein becomes dilated to resemble an extra cardiac chamber C P V = common pulmonary vein L A = left atrium L V = left ventricle

Endocardial Sclerosis

ENDOCARDIAL SCLEROSIS is characterized by elastic and collagenous thickening of the mural endocardium Usually the left ventricle is involved The left atrium may be involved as well The valves on the side of the heart affected may be stenotic but as a rule they are normal Two types of endocardial sclerosis may be distinguished on the basis of the appearance of the left ventricle the contracted type and the dilated type

The endocardial thickening of the left ventricle probably prevents normal excursion during the cardiac cycle of the ventricle and so causes a progressive impediment to pulmonary venous drainage The effects on the pulmonary circulation and right ventricle are similar to those which would be caused by mitral stenosis

Survival beyond infancy is uncommon

Endocardial Sclerosis, Contracted Type

IN THIS heart the left ventricular endocardium is thick and gray on the basis of elastic and collagenous thickening The left ventricle is of normal size but the left atrium is dilated and the right ventricle is hypertrophied



Fig 5 a—The left side of the heart The left ventricular endocardium is thick (see Figure 53a) The chamber s of normal size while the left atrium is dilated The mitral valve is normal



Fig 52b—The right ventricle is hypertrophied

History of the Patient

BOY 5 years old The parents had noted that fatigue and dyspnea occurred readily at the age of 2 years Since that date four episodes considered to be pneumonia which responded to antibiotics Two months before examination there was onset of cough fever dyspnea and edema of the extremities The fever responded to treatment but the cough persisted

Examination revealed marked cardiac enlargement rales over both lung fields hepatomegaly and dependent edema The second pulmonic sound was markedly accentuated The electrocardiogram revealed right axis deviation exaggerated P waves in leads I II and III Death from congestive heart failure

Principal Clinical Features of This Anomaly

1 Progressive dyspnea usually from an early age with eventual right heart failure
2 Accentuation of the second pulmonic sound
3 Evidence of right or left ventricular hypertrophy
4 Absence of the usual signs of acquired or congenital cardiac disease

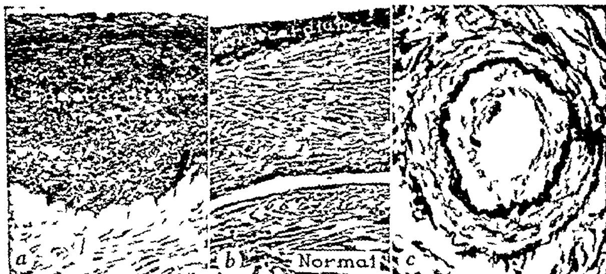


Fig 53—Photomicrographs of sections stained with Verhoeff's elastic tissue stain counterstained with van Gieson's connective tissue stain *a* Left ventricle of heart illustrated in Figure 5? Marked thickening of endocardium predominantly with elastic tissue Compare with normal in *b* (x 0) *b* Normal left ventricular endocardium and underlying myocardium from a 5 year old child (x 0) *c* Muscular pulmonary artery from the case described on page 3 Medial hypertrophy and intimal fibrous thickening (x340)

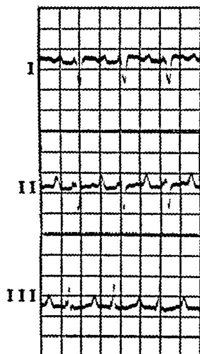




Fig 54—The electrocardiogram and the roentgenogram of the thorax from the patient whose heart is illustrated in Figure 52

Endocardial Sclerosis, Dilated Type

COMMONER than the contracted type of endocardial sclerosis is the dilated type illustrated on this page Usually there is a history of failure to gain weight and dyspnea and cardiac enlargement Cyanosis appears terminally and death results from congestive heart failure These cases have at times been referred to as examples of so called idiopathic cardiac hypertrophy

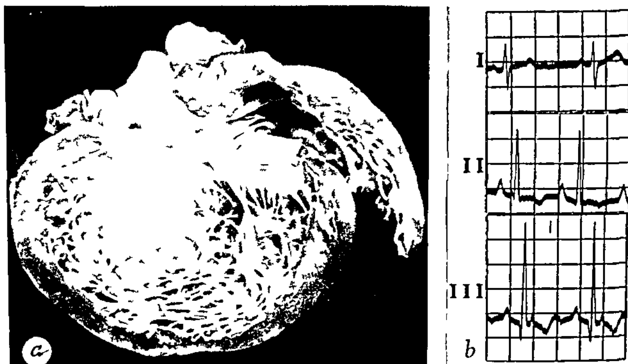


Fig 55—Endocardial sclerosis dilated type from a female 17 months old who appeared well until three weeks prior to admission when dyspnea and edema of the extremities appeared There was marked cardiac and hepatic enlargement No murmurs were heard Cyanosis appeared terminally *a* Left ventricular dilatation and hypertrophy The mural endocardium is thick opaque and gray *b* The electrocardiogram

Persistent Common Atrioventricular Canal

PERSISTENT common atrioventricular canal may be considered to appear in the complete form and in the partial form In the complete form mitral and tricuspid valves as such are not present There is only a common atrioventricular orifice and valve so that venous blood and arterial blood intermix as they traverse the atrioventricular canal There is also a defect in the atrial septum immediately above the common valve

In the partial form usually there is also a defect in the lower part of the atrial septum but the tricuspid valve is properly formed The mitral valve on the contrary shows a cleft in its anterior leaflet

In a rare case of the partial form the atrial septum is normally formed but there is a cleft in the mitral valve Those patients with a defect in the atrial septum suffer predominantly from the arteriovenous shunt while in the rarer type of the partial form with an intact atrial septum the functional disturbance is that of mitral insufficiency resulting from the deformity of the mitral valve

In the complete form survival beyond infancy is uncommon In the partial form some patients survive to adult life Mongolism occurs not infrequently in association with this congenital anomaly

Persistent Common Atrioventricular Canal, Complete Form

In this heart the atrioventricular canal is undivided and is guarded by a single valve composed of a large anterior cusp a large posterior cusp and smaller lateral cusps There is a defect in the lower portion of the atrial septum above the common atrioventricular canal





Fig 56a—Anterior view (model x1) (1) Dilated right atrium ( ) Enlarged right ventricle (3) Wide pulmonary trunk

Fig 56b—Interior of right atrium viewed from above (model x1) (1) Anterior cusp of common atrioventricular valve (2) Posterior cusp of common atrioventricular valve (3) Defect in lower portion of atrial septum

History of the Patient

Female 5 months old Clinical features of mongolism Transient cyanosis at birth Recurrent cyanosis during feeding and crying Frequent episodes of choking Patient admitted with pneumonia No murmurs heard Thoracic roentgenogram showed a large globular heart Lipoid pneumonia was found at necropsy

Principal Clinical Features of This Anomaly

1 Variable degree of cyanosis (the circulatory abnormality often resembles that in atrial septal defect)
2 Usually systolic murmur over midprecordium and apex
3 Roentgenologic aspects right and left ventricle equal in size and both enlarged
4 Electrocardiogram usually high voltage biphasic QRS complexes in standard and precordial leads



Fig 57a—Specimen from which models shown in Figure 56 were prepared Interior of right atrium and right ventricle

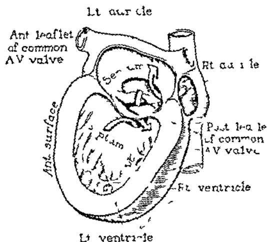


Fig 57b—Diagram of intracardiac circulation in persistent common atrioventricular canal



Fig 58—Thoracic roentgenogram of the patient whose heart is illustrated in Figures 56 and 57

Persistent Common Atrioventricular Canal, Complete Form

THE HEART illustrated on this page is from a female noncyanotic Mongolian idiot 12 months old Respira tory difficulty since birth Died suddenly en route to hospital

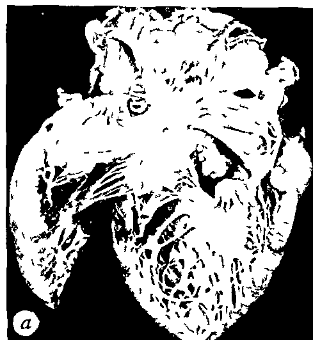




Fig 59—Persistent common atrioventricular valve When the heart is viewed in a conventional view there appears to be a cleft of the anterior leaflet of the mitral valve (*a*) and a similar condition of the septal leaflet of the tricuspid valve (*b*)



Fig 60—When the atrioventricular region is viewed from above it is apparent that there is neither mitral nor tricuspid valve as such but instead *there is one valve common to the two* sides of the heart (The atrial septum has been divided posterior to the defect in its lower portion and reflected forward)

Persistent Common Atrioventricular Canal, Partial Form in an Adult, Subacute Bacterial Endocarditis of Mitral Valve

A MAN 36 years old A cardiac murmur at 2 years of age Acyanotic until the development of bacterial endorcarditis and terminal congestive cardiac failure (From Rogers H M and Edwards J E *Am Heart J* 36 29 1948 with permission)

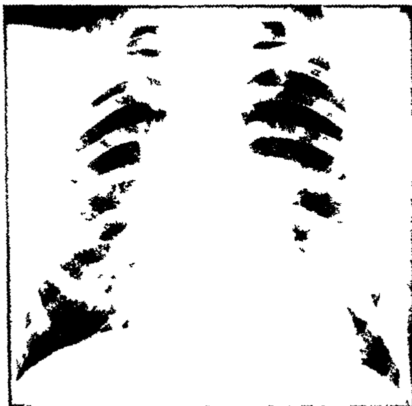


Fig 61—Roentgenogram of the thorax



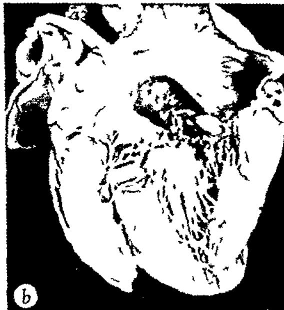


Fig 62—Partial form of common atrioventricular canal subacute bacterial endocarditis *a* Right side of heart Tricuspid valve is normal Defect lower part of atrial septum Vegetation on the mitral valve seen through defect *b* Left side of heart Cleft in anterior leaflet of mitral valve Vegetations on base of valve which forms lower limit of atrial septal defect

Persistent Common Atrioventricular Canal, Partial Form

In the complete form of persistent common atrioventricular canal there is one valve common to both sides of the heart. If one views the heart in a conventional manner, it gives the appearance of having clefts in the mitral and the tricuspid valves. There are, in fact, neither tricuspid nor mitral valves. In the partial form of the condition there are two atrioventricular valves, but the mitral valve is split into two halves, and usually, though not always, there is an associated defect in the lower part of the atrial septum.

The heart illustrated on this page is from a male 6 months old at the time of death. The clinical features had been characterized by mongolism and persistent cyanosis since birth. A precordial systolic murmur had been present. Death was due to bronchopneumonia.





Fig. 63—Partial form of common atrioventricular canal. In *a* is shown a normally formed tricuspid valve. Above it there is a crescent-shaped atrial septal defect similar to that seen in the complete form of the malformation. There is also present an atrial septal defect in the region of the foramen ovale. This is not an integral part of the malformation. In *b* is shown a cleft condition of the mitral valve. In all probability this valve was incompetent. (From Rogers, H. M., and Edwards, J. E.: *Am. Heart J.* 36: 28, 1948, with permission.)

Persistent Common Atrioventricular Canal, Partial Form

(Intact Atrial Septum Mitral Insufficiency)

THIS HEART represents a partial form of common atrioventricular canal In it the atrial septum is intact and the tricuspid valve is normal There is a cleft in the anterior leaflet of the mitral valve allowing mitral insufficiency (Specimen clinical history electrocardiogram and roentgenogram submitted by Drs Charles Hollis and Leslie S Jolliffe Reproduced with their p[illegible])

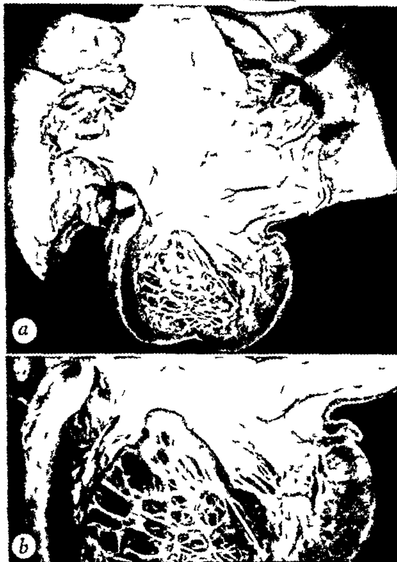


Fig 64a—The left side of the heart Cleft in anterior leaflet of mitral valve Intact atrial septum Dilatation of left atrium and left ventricle

Fig 64b—Detail of mitral valve showing cleft of anterior leaflet

History of the Patient

BOY 8 10/12 years old Apparently well though underdeveloped during infancy Recurrent asthma like attacks without cyanosis began during second year At the age of 3 years cardiac failure developed but the heart responded to digitalis Pulmonary symptoms well controlled with prophylactic sulfonamides At 5 years findings were retarded growth precocity slight clubbing of the fingers cardiac enlargement with prominence of the left side of the thorax loud harsh systolic and diastolic murmurs over left side of thorax and back thrill at apex blood pressure 90/60 episodes of paroxysmal tachycardia At 7 years cardiac catheterization (permission of Dr R J Bing) revealed no shunts but pressure in right ventricle and left pulmonary artery elevated to 48/0 and 48/22 respectively Systemic arterial blood 92 per cent oxygen saturation at rest and 91 per cent during exercise Erythrocytes 4 880 000 hemoglobin 15 gm From the age of 7 years on progressive dyspnea and eventually cardiac failure

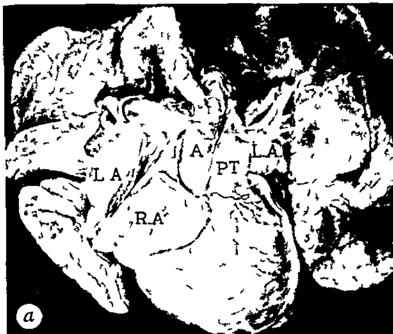




Fig 65—From the case described on page 41 *a* Anterior view of exterior of thoracic organs Dilatation of pulmonary trunk (P T) The dilated left atrium (L A) extends to the right of the right atrium (R A) A = aorta *b* Posterior view of thoracic organs Horizontal position of major bronchi secondary to effects of dilated left atrium

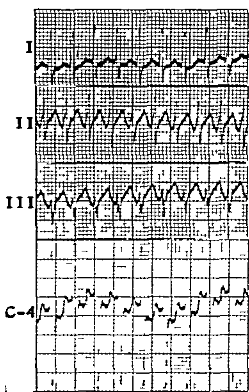




Fig 66—The electrocardiogram and roentgenogram of the thorax of the patient from whom specimens are illustrated in Figures 64 and 65

Atrial Septal Defect

ATRIAL septal defects are among the commoner types of congenital cardiac anomalies seen in adult life The usual form of atrial septal defect is a valvular incompetence of the foramen ovale for one or more of three reasons the valve of the foramen may be short the foramen may be unusually large or there may be perforations in the valve of the foramen ovale The defect allows oxygenated blood to flow from the left atrium to the right atrium This shunt results in recirculation of oxygenated blood through the lungs

If at all appreciable the shunt causes dilatation and hypertrophy of the right ventricle enlargement of the right atrium and enlargement of the pulmonary trunk and its branches Although the pressure in the pulmonary artery may be moderately elevated it is unusual to find secondary changes in the small muscular arteries and arterioles As long as the shunt is arteriovenous there is no cyanosis

Symptoms may be minimal or absent for years In other cases congestive heart failure develops sudden death may occur Occasionally an atrial septal defect is associated with acquired rheumatic mitral stenosis constituting the so-called Lutembacher syndrome A rare complication is brain abscess The escape of an embolus through an atrial septal defect results in the phenomenon described as paradoxical *embolism*

Atrial Septal Defect

IN THESE two hearts the anomaly is an atrial septal defect in the form of a patent foramen ovale As a consequence of the left to right shunt there is enlargement of the right atrium and right ventricle and of the pulmonary trunk

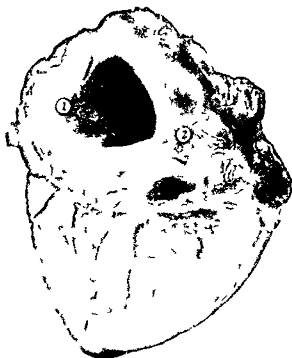


Fig 67a—Interior of right atrium viewed from above (model x1) (1) Margin of large defect in atrial septum (2) Dilated right atrium

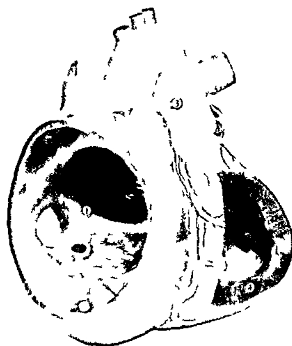


Fig 67b—Heart viewed from right (half size model) (1) Margin of large defect in atrial septum (2) Interior of dilated right atrium (3) Dilated pulmonary trunk (4) Hypertrophied wall of enlarged right ventricle

History of These Patients

FEMALE 5 years old (Fig 67*a*) Periodic cyanosis beginning at 1 year Pneumonia at 3 years No murmurs heard at any time Intermittent auricular flutter Patient died with congestive heart failure

MAN 75 years old (Fig 67*b*) Recurring congestive failure for 11 years Right hemiplegia at 64 years Loud precordial systolic murmur Thoracic roentgenogram (Fig 69) shows tremendous enlargement of heart and electrocardiogram shows right axis deviation Patient died with congestive heart failure

Principal Clinical Features of This Anomaly

1 Characteristically no cyanosis 2 Systolic murmur usually present not diagnostic in type Accentuated second sound at pulmonary area occasionally a soft diastolic murmur of pulmonary insufficiency 3 A major defect may be present for many years without cardiac symptoms Unexpected sudden death occasionally occurs 4 High oxygen saturation of blood in right atrium as compared to that of blood in venae cavae greatly increased pulmonary blood flow (cardiac catheterization) 5 Roentgenologic aspects marked enlargement of right atrium right ventricle and pulmonary artery Accentuated hilar pulsations Occasionally aneurysmal dilatation of pulmonary artery 6 Electrocardiogram right axis deviation Right ventricular hypertrophy pattern or delayed conduction of the right ventricle (right bundle branch block) Auricular fibrillation not infrequently present



Fig 68a—Specimen from which the model shown in Figure 67a was prepared Interior of right atrium



Fig 68b—Specimen from which the model shown in Figure 67b was prepared Interior of the left atrium and mitral valve

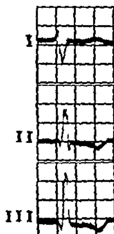
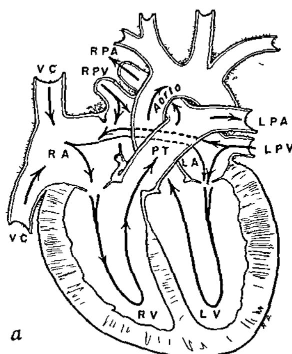
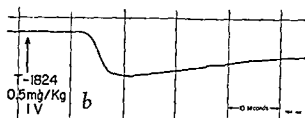


Fig 69—Electrocardiogram and thoracic roentgenogram of the patient whose heart is illustrated in Figures 67b and 68b

Atrial Septal Defect, Clinical Case

MAN 24 years old No symptoms during childhood Murmur at 9 years At age 24 years mild effort dyspnea only Systolic murmur over the pulmonary area and a prolonged pulmonary diastolic murmur





Synopsis of Significant Catheterization Data

	Pressure mm Hg	O saturation, per cent
Superior vena cava	11/6	77 5
Inferior vena cava	10/5	80 5
Right atrium	12/6	90 5
Right ventricle	35/10	90 0
Pulmonary artery	31/18	90 0
Radial artery	140/78	98 0

Flow values liters/min /M² *Systemic 3 9 Pulmonary 8 0*

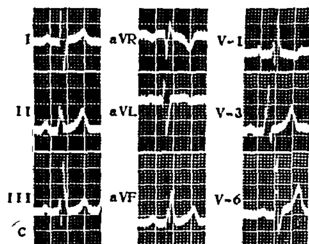


Fig 70a—(uppe left)—Intracardiac circulation in atrial septal defect *b (upper right)* The dye dilution curve *c (left)* The electrocardiogram *d* and *e (below)* The anteroposterior and lateral roentgenograms of the thorax



Representative Dye-Dilution Curves Recorded by Ear Oximeter

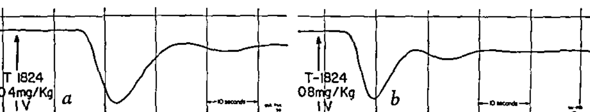


Fig 71a and b—The oximeter has been utilized to record the arterial blood concentrations of a dye (T1824) after a single rapid intravenous injection of this dye The graphic records so obtained are usually analyzed to give measurements of the circulation time cardiac output and blood volume In these records increasing concentration of the dye is represented by a downward deflection In the curve from a normal person (a) it may be noted that the appearance time of the dye (circulation time) is 12 seconds and there is a rapid increase in the dye concentration followed by a rapid decrease Detailed discussion of normal and abnormal curves is available in the article by Nicholson J W et al (J Lab & Clin Med 37 353 1951) The dye dilution curve in b was obtained from a child with isolated pulmonary stenosis It is of normal contour which is the characteristic finding in patients with this lesion in the absence of heart failure The arm to ear circulation time (6 seconds) is within the normal range for children

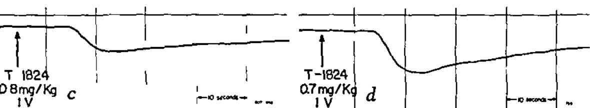


Fig 71c and d—The records shown above were obtained from 2 patients having large left to right shunts c the curve from a patient with partial anomalous pulmonary venous drainage d the curve from a patient with a ventricular septal defect the diagnosis in each instance being made following cardiac catheterization The characteristic feature of these curves having diagnostic value is the very gradual decrease in concentration of the dye following the initial peak In patients with left to right shunts the dyed blood is recirculated through the defect and pulmonary circulation so that initial clearance of dye from the heart and lungs is markedly prolonged It is to be noted that the appearance time of the dye (circulation time) is normal It will be apparent that the diagnosis of left to right shunt may be indicated by these curves but the anatomic site of the shunt cannot be designated A curve similar to these is obtained in patients with atrial septal defects (see Figure 70b page 46) Shunts of magnitudes of less than 20 per cent may not be discernible by this method of investigation

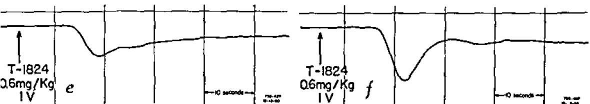


Fig 71e and f—The records shown above were obtained from a patient with a patent ductus arteriosus before (e) and after (f) its surgical closure the latter record being essentially normal The record obtained before operation shows the prolonged disappearance slope characteristic of left to right shunts Sometimes as in this case there appears to be a recognizable break in the smooth contour of the curve following its apex related probably to the first pulmonary recirculation of the dye and suggesting the diagnosis of patent ductus arteriosus as opposed to an atrial septal defect From the comparison of the preoperative and postoperative curves assuming equivalent sensitivity of the recordings it is evident that the initial peak of dye concentration may be much diminished in left to right shunts The abnormal curves obtained from patients with left to right shunts may be compared with that obtained from a patient with a right to left (venous arterial) shunt by referring to Figure 133 page 84

Patent Foramen Ovale, Anatomic and Functional Studies





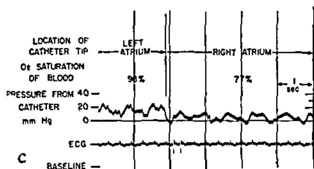


Fig 72*a*—Probe patent foramen ovale (containing probe) viewed from the left *b* Probe patent foramen ovale (containing probe) viewed from the right *c* Catheterization data when catheter had passed through probe patent foramen ovale into the left atrium and after it had returned to the right atrium From a male 3 months old who suffered from a patent ductus arteriosus and cardiac failure

Atrial Septal Defects, Anatomic Studies



Fig 73*a*—Left side of heart Atrial septal defect caused by (1) an abnormally short valve of the foramen ovale and (2) multiple perforations in the valve



Fig 73*b*—Right side of heart Close relationship between the inferior vena cava (containing probe) and the atrial septal defect

Probe-Patent Foramen Ovale, Paradoxical Embolus

THE PATIENT was a 64 year old man who died as a result of pulmonary embolism A paradoxical embolus also was found lodged in the foramen ovale There was probably no shunt through the probe patent foramen ovale until pressure in the right atrium became elevated as a result of pulmonary embolism Then further embolism to the right atrium allowed the paradoxical embolism to occur



Fig 74a—Right side of heart A long thrombus evidently originating in a vein is engaged in the opening of a probe patent foramen ovale (between arrows)



Fig 74b—Left side of heart The left extremity of the thrombus seen in *a* presents in the left atrium through the opening of a probe patent foramen ovale (circle)

Atrial Septal Defect, Several Positions of Cardiac Catheter

From a patient 35 years old (From Burchell H B *J Iowa M Soc* *38* 364 1948 with permission)



Fig 75a—A catheter is passed through the atrial septal defect and its tip lies in a right pulmonary vein *b* The catheter has passed through the atrial septal defect and its tip lies in a left pulmonary vein *c* The catheter has passed through the atrial septal defect and mitral valve Its tip lies in the left ventricle Blood completely saturated with oxygen was obtained from each of the three positions of the catheter illustrated

Atrial Septal Defect Complicated by Abscess of the Brain

The patient was a 23 year old woman (From Gates E M *et al* *Proc Staff Meet Mayo Clinic* 22 401 1947 with permission)



Fig 76*a*—The atrial septal defect is seen from the left side



Fig 76*b*—A solitary abscess in the right parietal lobe which involved the right parietal and occipital lobe Culture of the abscess revealed Actinomyces bovis No inflammatory disease of the heart was present

Lutembacher's Syndrome

THE PATIENT was a man 50 years old Paroxysmal auricular fibrillation for years Admitted with congestive failure auricular fibrillation cardiac enlargement Murmurs of mitral stenosis Sudden death during gradual improvement of cardiac failure

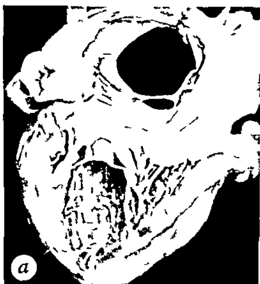


Fig 77*a*—Left side of heart Atrial septal defect Mitral stenosis (From Edwards J E *Postgrad Med* 3 377 1948 with permission)

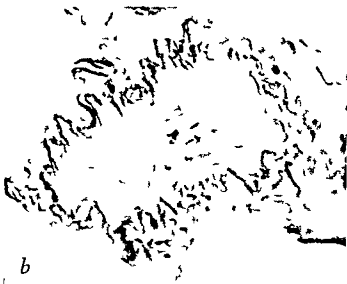


Fig 77*b*—A pulmonary arteriole from this case showing marked intimal fibrous thickening causing luminal narrowing (Verhoeff's elastic tissue stain, counterstained with van Gieson's connective tissue stain) Lesions of this character were relatively widespread in the lungs Such changes are only rarely found in a widespread manner in uncomplicated atrial septal defect

Anomalous Drainage of Pulmonary Veins

PULMONARY veins may drain anomalously into the right atrium or one of its tributary veins instead of into the left atrium Such anomalous venous drainage may be partial or complete In partial anomalous drainage some of the pulmonary veins communicate normally with the left atrium others communicate anomalously with the right atrium or one of its tributary veins In complete anomalous drainage all the pulmonary veins drain into the right atrium or one of its tributary veins none drain into the left atrium In the latter anomaly an atrial septal defect is present which is the only route whereby blood can reach the left side of the heart for distribution to the systemic circulation

In partial anomalous venous drainage there is pulmonary recirculation of oxygenated blood and the condition has functional characteristics similar to those of atrial septal defect No cyanosis is present Survival to adult life usually occurs

In complete anomalous drainage of the pulmonary veins there is not only an arteriovenous but also a venous arterial shunt In this condition the arterial blood is incompletely saturated with oxygen and cyanosis is usually evident Death during early infancy is common

Partial Anomalous Drainage of Pulmonary Veins

In figure 78 is depicted diagrammatically partial anomalous drainage of the pulmonary veins The drainage from the right lung is into the superior vena cava while blood from the left lung drains normally into the left atrium

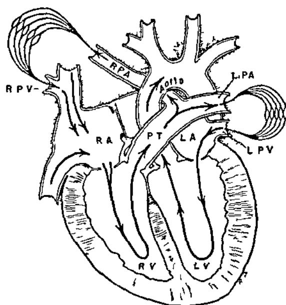


Fig 78—Anomalous drainage of blood from right lung into superior vena cava

History of the Patient

Girl 9 years old with cardiac murmur known to be present since birth Small but well developed Good exercise tolerance Never any cyanosis Coarse systolic murmur over the base transmitted to the interscapular region Roentgenoscopy showed some cardiac enlargement with prominent shadows of pulmonary trunk and increased pulsations of pulmonary vessels Electrocardiogram showed evidence of right ventricular hypertrophy Hemoglobin 12 2 gm per 100 cc Cardiac catheterization showed pulmonary vein entering right atrium (Fig 80) Oxygen saturation of hemoglobin at this site was 98 per cent

Principal Clinical Features of This Anomaly

1 Tachycardia no cyanosis shunt is arteriovenous

2 Variable systolic basal murmur accentuation of second pulmonic sound symptoms may be mild or absent

3 Abnormally high oxygen saturation of hemoglobin of blood in right atrium or superior vena cava If catheter passes directly from superior vena cava or from right atrium into a pulmonary vein a positive diagnosis is established

4 Roentgenologic aspects right ventricular enlargement with prominent pulmonary artery and increased hilar pulsations

5 Electrocardiogram evidence of right ventricular hypertrophy

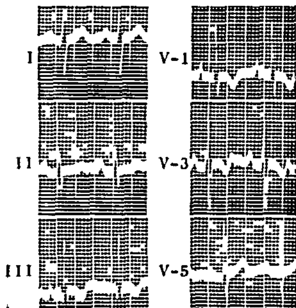




Fig 79—The electrocardiogram and the roentgenogram of the tho ax of the patient whose history appears on p ge 5?

Synopsis of Catheterization Data

	Pressure mm Hg	O satu ation per cent
Supe ior vena cave	5/3	78
Infer or en ca a	5/3	83
Right atrium	8/4	89
R ght ventricle	25/5	90
Pulmonary artery	3/1	91
Right pulm nary ein	6/4	98



Fig 80—Roentgenogram showi g that the card ac catheter h s passed to the right of the cardiac bordc The blood obt ined from th s site was 98 per cent sat ated w th oxy gen i d cating that the catheter l es n a pulmon ry vein

Complete Anomalous Drainage of Pulmonary Veins Into Superior Vena Cava, Atresia of Lower Portion of Common Pulmonary Vein

In this heart all the pulmonary veins drain into a common vessel The latter is connected with the left atrium by an atretic strand The only exit of blood from the common vessel is by way of an anomalous connection with the superior vena cava

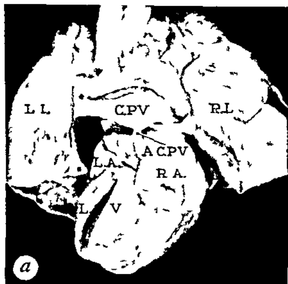


Fig 81a—The heart and lungs from behind The pulmonary veins enter a common vessel the common pulmonary vein (C P V) An atretic strand (A C P V) extends from the common pulmonary vein to the left atrium (L A) but there is no channel connecting directly the common pulmonary vein with the heart

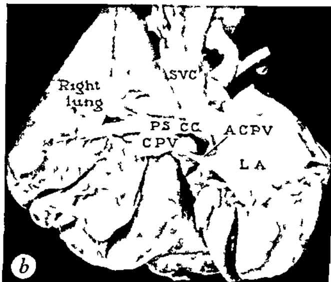


Fig 81b—Heart and right lung from right anterior position From the common pulmonary vein (C P V) an anomalous vein leads to the superior vena cava (S V C) R A = right atrium L V = left ventricle L L = left lung R L = right lung P S C C = anomalous pulmonary venous superior vena caval connection

History of the Patient

Boy 7 weeks old cyanotic at birth but improved in oxygen Subsequently cyanosis occurred during crying and feeding Infant was flabby neck was tilted to the left respirations rapid with edema about the neck and lower jaw No cardiac murmurs Roentgenogram of the thorax showed marked cardiac enlargement upper vascular shadow broad Electrocardiogram showed right ventricular hypertrophy Esophagrams revealed no obstruction or deformity Erythrocytes 3 670 000 Hemoglobin 12 2 gm per 100 cc of blood

Cyanosis became more pronounced in spite of constant administration of oxygen Edema of neck increased extending to the face and scalp Respirations became more rapid and noisy (From Edwards J E *et al* *Arch Path* 51 446 1951 with permission)

Principal Clinical Features of This Anomaly

1 Some degree of cyanosis is usually present from birth
2 Typically no murmurs
3 Cardiac enlargement (right side) with evidence of pulmonary congestion
4 Electrocardiographic evidence of right ventricular hypertrophy Usually death in early infancy but rarely patients survive to adolescence and adulthood

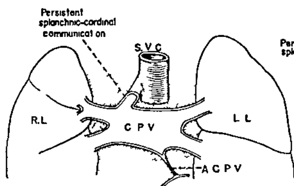


Fig 82—From the case illustrated in Figure 81 The pulmonary veins enter a common chamber (C.P.V.) which in turn is connected by an anomalous channel to the superior vena cava (S.V.C.) A.C.P.V. = a strand of tissue representing the atretic lower portion of the common pulmonary vein of the embryo

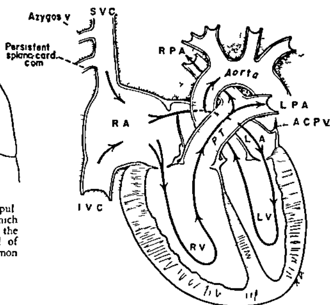


Fig 83—The intracardiac circulation in complete anomalous drainage of the pulmonary veins

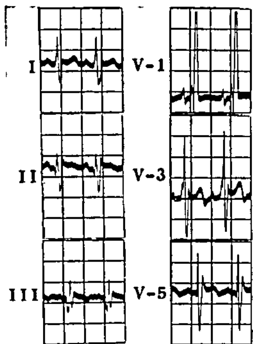




Fig 84—The electrocardiogram and the roentgenogram of the thorax of the patient whose heart and lungs are illustrated in Figures 81 83

Complete Anomalous Drainage of Pulmonary Veins into Ductus Venosus

ILLUSTRATED on this page is the case of a male infant who died at the age of 9 days following a period of cyanosis but with no abnormal auscultatory signs The entire pulmonary venous drainage was into the ductus venosus (From Edwards J E and DuShane J W *Arch Path* 49 517 1950 with permission)



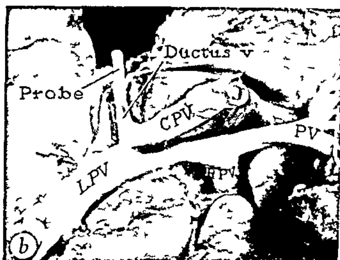
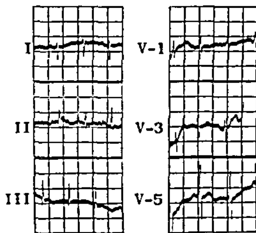




Fig 85—(above)—a The lungs from the front The heart has been deflected to the right The pulmonary veins join to form a common pulmonary venous trunk (C P V) which does not enter the heart It descends *in company with the esophagus through the diaphragm b* After reaching the abdominal cavity the common pulmonary venous trunk (C P V) enters the ductus venosus (Ductus) L P V = left portal vein R P V = right portal vein P V = portal vein The upper end of the probe represents the junction of the ductus venosus with the left *hepatic vein*

Fig 86—(left)—The left side of the heart There is a patent foramen ovale which represents the only route by which the left atrium could receive blood

Fig 87—(below)—The electrocardiogram and the roentgenogram of the thorax





Congenital Pulmonary Arteriovenous Fistula

THIS anomaly as the name implies is a direct communication between the pulmonary arterial system and the pulmonary venous system This allows venous unsaturated blood to enter the pulmonary veins and thus the systemic circulation When the fistula is large and there is sufficient venous blood entering the systemic circulation cyanosis clubbing of digits and secondary polycythemia are manifest Cardiac murmurs are characteristically absent but a bruit over either lung field with a corresponding density in the roentgenogram of the chest are valuable clues in the diagnosis

Only one lobe may be involved in which case the lesion or the affected lobe can be surgically removed When however multiple lobes are involved surgical treatment may not be possible Patients with pulmonary arteriovenous fistula frequently reach adult life Telangiectasia of the skin and mucous membranes is relatively common Brain abscess may occur as a complication of this congenital anomaly as is true in any of the anomalies which allow venous blood to enter the systemic circulation without traversing the pulmonary capillary bed

Congenital Pulmonary Arteriovenous Fistula

ILLUSTRATED are the hands of the patient before and after a lobectomy for pulmonary arteriovenous fistula

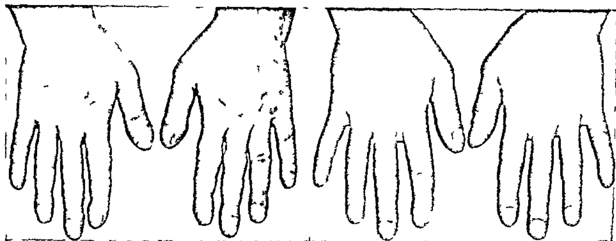


Fig 89 —Hands of the patient *a* Taken June 14 1946 before lobectomy for arteriovenous fistula *b* Taken on March 1 1949 2½ years after the operation Clubbing of fingers has disappeared as has cyanosis

History of the Patient

MAN 20 years old Normal development No disability in preschool years At the age of 8 years cyanosis of the lips first noted Three years prior to examination dyspnea on climbing stairs Examination revealed cyanosis clubbing of the fingers and toes and enlargement of the wrist knee and ankle joints Heart was not appreciably enlarged No cardiac murmurs Over a small area below the right nipple a late systolic bruit which varied in intensity with phase of respiration Loudest in midinspiration Roentgenogram showed nodular shadow in the right lung and a normal cardiac contour Nodular shadow was noted to pulsate on roentgenoscopy The electrocardiogram was normal

Hemoglobin was 24 1 gm erythrocytes 7 590 000 hematocrit 82 per cent blood volume 172 cc per kilogram

Resection of the middle lobe of the right lung successfully carried out When seen 3 years later patient was well Hemoglobin 14 3 gm erythrocytes 5 120 000 hematocrit 49 per cent electrocardiogram normal (From Burchell H B and Clagett O T *Am Heart J* *34* 151 1947 with permission)

Principal Clinical Features of This Anomaly

1 Cyanosis becoming apparent in youth and increasing in severity
2 Polycythemia and clubbing of the fingers and toes
3 Absence of cardiac murmurs and other signs of primary cardiac disease
4 Bruit over some part of the lung fields in some cases This may be a continuous type of murmur or a systolic murmur
5 Small cutaneous capillary hemangiomas occur in about half of the cases
6 Cardiac enlargement occurs in the minority of patients

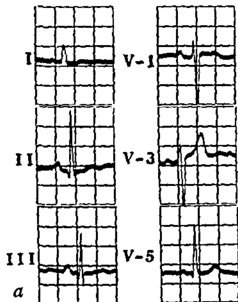


Fig 89a—The electrocardiogram before operation

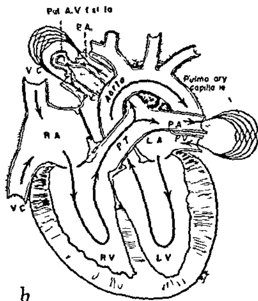


Fig 89b—The circulation in pulmonary arteriovenous fistula

Fig 90—The roentgenogram of the thorax before (6-1-46) *(left)* and after (9-16-46) *(right)* right middle lobectomy for pulmonary arteriovenous fistula From patient whose hands are illustrated in Figure 88 whose electrocardiogram appears in Figure 89a and whose case history is described on page 58

Pulmonary Arteriovenous Fistula, Abscess of Brain

A WOMAN 43 years old developed amnesic episodes with confusion in 1944 Visual field examination revealed left homonymous hemianopsia Thoracic roentgenogram reported as negative A right temporal craniotomy revealed a multiloculated brain abscess which was evacuated and enucleated The patient made a complete recovery

In 1950 a routine roentgenologic examination of the thorax revealed an irregular shadow in the left hilar region Results of bronchoscopic examination were negative An exploratory thoracotomy revealed enlargement of the left pulmonary artery and vein Further dissection proved the mass to be vascular in nature and a left lower lobectomy was performed Lobe contained an arteriovenous fistula 3 cm in diameter Progress has been satisfactory since





Fig 91a—(above)—Abscess of brain enucleated in 1944 *b (right)* Roentgenogram of thorax in 1950 revealing shadow of pulmonary arteriovenous fistula in hilus of left lung

Ventricular Septal Defect

VENTRICULAR septal defects occur either in the membranous or in the muscular portion of the ventricular septum In either case a shunt of blood from the left ventricle to the right ventricle occurs with each systolic ejection and increases the load of work of both ventricles The postnatal period seems to be a hazardous one for some infants with ventricular septal defect and death during the first year of life is common Patients who survive this period however may live many years without disability Those who reach adulthood may show little or no cardiac enlargement and the electrocardiogram may be normal The so called Roger murmur is an important diagnostic sign in this anomaly

Bacterial endocarditis is a common complication in patients who survive infancy

Certain variations of ventricular septal defect occur Occasionally a ventricular septal defect will be associated with a deformity of the adjacent aortic valve producing aortic insufficiency and in rare instances the ventricular septal defect is so disposed as to establish a communication between the left ventricle and the right atrium

Ventricular Septal Defect

Two types of ventricular septal defects are illustrated by these two hearts In one the defect is in the muscular portion of the ventricular septum In the other the defect involves the membranous portion The latter type is the commoner



Fig 9 —Interior of left ventricle (model x1) (1) Margin of defect in muscular portion of ventricular septum (2) Intact membranous ventricular septum



Fig 93—Interior of left ventricle (half size model) (1) Margin of defect in membranous portion of ventricular septum ( ) Wide pulmonary trunk (3) Aortic orifice

History of These Patients

Boy 5½ months old (Fig 92) had had feeding difficulty since birth with poor development A loud systolic murmur was maximal in third left interspace Dyspnea present for two months Roentgenogram (Fig 96) shows moderate enlargement of heart Progressive failure with terminal cyanosis

Woman first seen at the age of 32 years (Fig 93) had a loud harsh systolic murmur over entire precordium accentuated p 2 mild exertional dyspnea enlarged heart She was readmitted at 54 years because of fracture of femur Slight cyanosis and mild congestive heart failure were found She died of pulmonary embolism

Principal Clinical Features of This Anomaly

1 Loud systolic murmur maximal in third left interspace associated thrill
2 No cyanosis
3 High oxygen saturation of blood in right ventricle as compared to that of blood in right atrium (cardiac catheterization)
4 High incidence of subacute bacterial endocarditis
5 Roentgenologic aspects heart is normal in size and contour in most instances
6 Electrocardiogram usually normal (congenital heart block which is rare usually occurs with this malformation)



Fig 94—Inter or of the left ventricle Specimen from which the model shown in Figure 9 was prepared



Fig 95—Interior of the left ventricle Specimen from which the model shown in Figure 93 was prepared

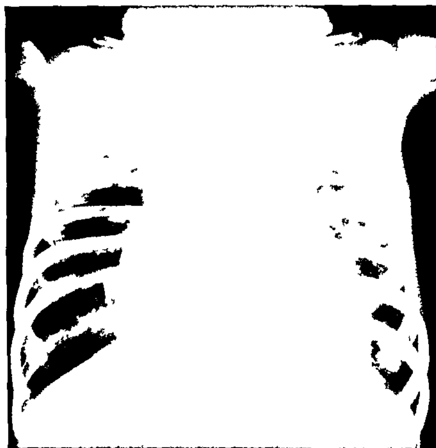
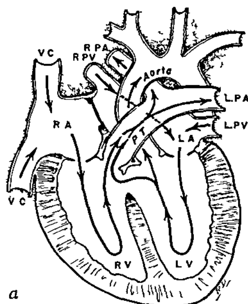


Fig)6—Thorac c roentgenogram of the pat ent wl ose heart is illustrated n F gures 92 and 94

Ventricular Septal Defect, Clinical Case

(Chosen to Demonstrate More Typical Clinical Picture)

MAN 29 years old Cardiac murmur known since childhood Asymptomatic Loud systolic murmur associated with thrill maximal intensity over fourth left interspace and obliterating both the first and second cardiac sounds Hemoglobin 14.3 gm per 100 cc



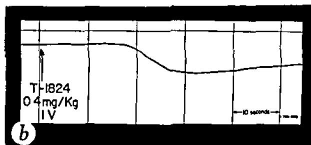


Fig 97a—The intracardiac circulation in ventricular septal defect (From Edwards J E *Arch Surg* 61 1103 1950 with permission) *b* The dye dilution curve

Synopsis of Significant Catheterization Data

	Pressure mm Hg	O saturation per cent
Superior vena cava	3/1	69.5
Inferior vena cava	4/1	71.5
Right atrium	3/1	71.0
Right ventricle	25/4	78.0 86.5
Pulmonary artery	18/9	81.5
Radial artery	130/71	98.5

Flow values liters/min /M Systemic 2.2 Pulmonary 5.0

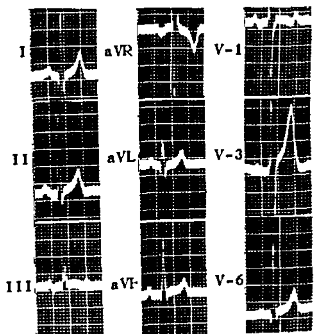




Fig 98—The electrocardiogram and the roentgenogram of the thorax

Ventricular Septal Defect in an Infant

Boy 5 months old at death Systolic murmur and thrill over entire precordium first noted at 5 days of age Paroxysmal coughing and slight cyanosis at 2 months Cardiac enlargement Pulmonary congestion and death





Fig 99—(left)—*a* Right ventricle Probe lies in right ventricular opening of a muscular ventricular septal defect Right ventricular hypertrophy *b* Left atrium and ventricle Enlargement of left atrium associated with secondary endocardial sclerosis Left ventricle hypertrophied and somewhat dilated Defect not visible in this view

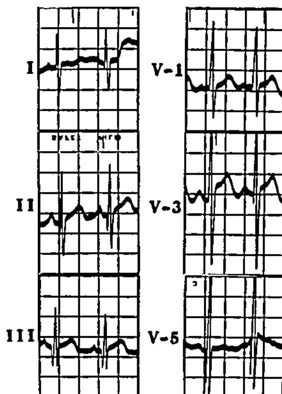
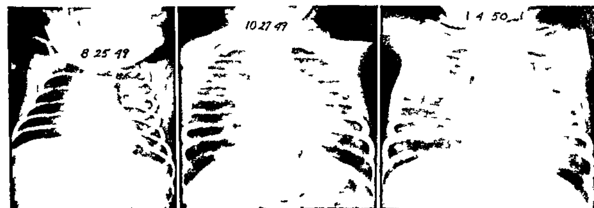


Fig 100—(above)—The electrocardiogram at 9 weeks of age

Fig 101—(below)—Roentgenograms of thorax taken at the ages of 5 days 9 weeks and 4½ months respectively showing progressive enlargement of heart Compare with roentgenograms of case of patent ductus arteriosus described on page 11 (Figure 190c)



Ventricular Septal Defect Associated with Deformity of the Aortic Valve and Aortic Insufficiency

A BOY 17 years old A machinery like precordial murmur (Specimen of Dr John C Henthorne) The association of aortic insufficiency with ventricular septal defect may yield signs suggesting patent ductus arteriosus

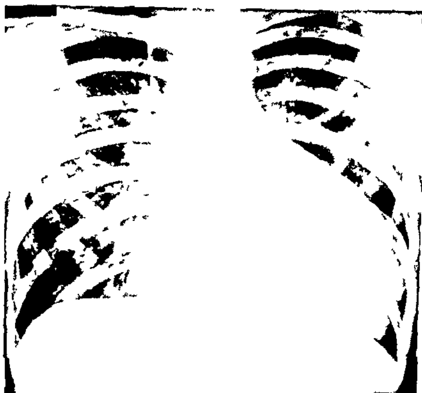


Fig 102—(above)—The roentgenogram of the thorax



Fig 103a—The left ventricle Hypertrophy and dilatation The upper margin of a ventricular septal defect involving the membranous portion of the septum is formed by the right and posterior aortic leaflets which are deformed



Fig 103b—Close up view of aortic valve The deformity of the aortic valve probably accounted for aortic insufficiency This together with the murmur of the ventricular septal defect created the murmur described

Ventricular Septal Defect with Communication Between Left Ventricle and Right Atrium

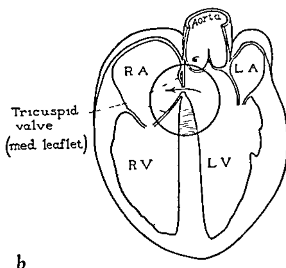



Fig 104—From a girl 5 years old who died of bacterial endocarditis of the edges of the ventricular septal defect and the anterior wall of the right ventricle (From Perry E L *et al* *Proc Staff Meet Mayo Clin* 24 198 1949 with permission) *a* A crescent shaped defect (point of arrow) exists in the tricuspid valve This represents a so-called double orifice of the tricuspid valve The edges of the orifice were adherent to the edges of a membranous ventricular septal defect allowing communication between the left ventricle and the right atrium as illustrated in *b* (In illustration *a* shadow behind defect represents area from which tissue removed for section)

Muscular Ventricular Septal Defect in an Adult

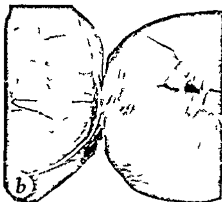



Fig 105—*a* Interior of left ventricle showing healed muscular ventricular septal defect in a man 65 years old The superior and inferior edges of the defect are almost in apposition suggesting that in this type of septal defect postnatal closure may possibly occur *b* Photomicrograph through defect illustrated in *a* The edges of the defect are fused There is no evidence of underlying myocardial disease that could explain the development of the defect on an acquired rather than a congenital basis (From Edwards J E In Gould S E *Pathology of the Heart* Charles C Thomas Publisher Springfield Ill no 1953 pp 266-503)

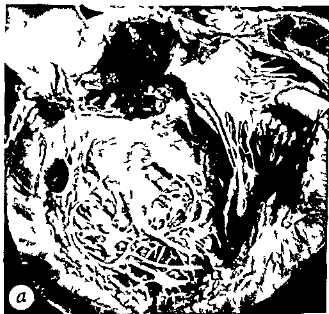
Ventricular Septal Defect with Biventricular Origin of Pulmonary Trunk





Fig 106—Heart from a male 1 week old Specimen submitted by Dr James R Dawson Jr *a* Right ventricle and pulmonary valve Probe placed in left ventricle extends into overlying pulmonary trunk *b* Left ventricle and aorta Probe lies in left ventricle and disappears through defect into the pulmonary trunk

Bacterial Endocarditis Complicating Ventricular Septal Defect



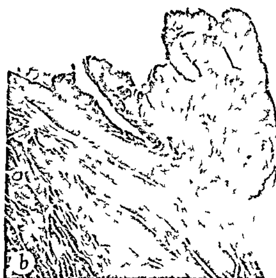


Fig 107—From a man [illegible]1 years old with bacterial endocarditis complicating a defect of the membranous portion of the ventricular septum *a* The left ventricle showing vegetations of bacterial endocarditis involving the aortic valve and endocardium of adjacent septal defect (From Edwards J E *Postgrad Med* 3 3[illegible]7 1948 with permission) *b* Section through edge of the septal defect showing vegetations of bacterial endocarditis deposited on the endocardial surface (hematoxylin and eosin x10) The traumatic effect of blood flow through a ventricular septal defect is probably the factor responsible for the high incidence of bacterial endocarditis in patients with ventricular septal defect who survive infancy

Eisenmenger Complex

(Ventricular Septal Defect with Biventricular Origin of the Aorta)

In the Eisenmenger complex the heart varies little from that in a simple defect of the membranous portion of the ventricular septum Because the aorta arises above the ventricular septal defect the right ventricle shares with the left the function of propelling blood into the aorta There is no pulmonic stenosis yet the pressure in the two ventricles is similar being at systemic levels Pressure in the pulmonary arterial system is equally high This feature depends on changes recognizable morphologically within the arteries of the pulmonary vascular bed Were it not for the obstructing effects resulting from these vascular changes the pulmonary system where the pressure normally is low would be flooded and a balanced circulation could thus not be maintained The small arteries in the lungs in the Eisenmenger complex resemble closely those of the normal fetus in which the increased resistance to pulmonary blood flow makes it possible for the blood to be shunted into the aorta through the functioning ductu arteriosus

Eisenmenger Complex, Clinical Case

MAN 43 years old Cyanosis since birth Periodic hemoptysis since age of 18 Reduced exercise tolerance Systolic and diastolic murmurs maximal over left third interspace Erythrocytes 6 970 000

Synopsis of Catheterization Data

	Pressure mm Hg	O saturation per cent	Simultaneous ear oximeter reading O saturation per cent
Superior vena cava	22/16	38	61
Inferior vena cava	2/15	39	68
Right atrium	25/10	40	69
Right ventricle	117/19	39	60
Pulmonary artery	129/43	42	62
Radial artery	160/75	62	60

Fig 11—Roentgenogram of thorax of the patient whose cardiac catheterization data and electrocardiogram are reproduced on this page

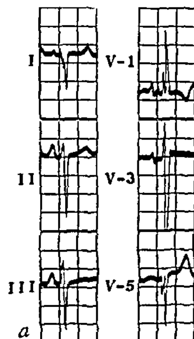


Fig 113a—The electrocardiogram

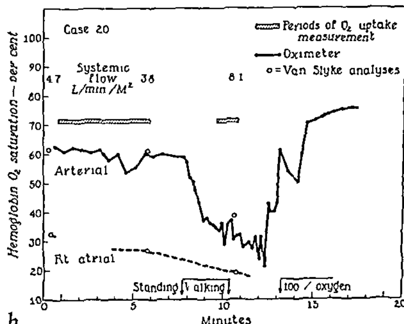


Fig 113b—Graphic demonstration of the effect of walking on oxygen saturation of arterial and venous blood and systemic blood flow (From Burchell H B *et al* *Circulation* 1 401 1950 with permission)

Eisenmenger Complex, Pulmonary Vasculature

THE HIGH pulmonary blood pressure characteristic of the Eisenmenger complex depends on a high resistance to blood flow through the pulmonary vascular bed On the basis of anatomic studies the zone of high resistance to flow seems to lie at the level of the intrapulmonary muscular arteries As in the normal fetus these vessels show thick mediae and correspondingly narrow lumina Later intimal fibrous thickening may be superimposed

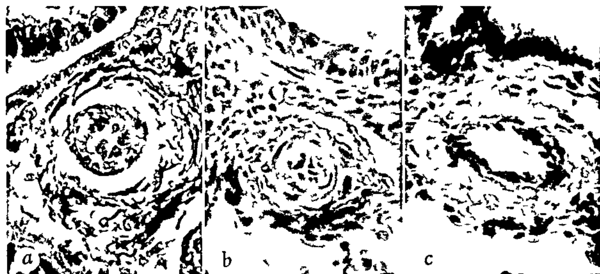


Fig 114—Photomicrographs of intrapulmonary muscular arterie (x450) *a* In a normal 5 month old fetus the lumen is narrow and the media thick *b* In the case of the Eisenmenger complex ill strated in Figures 108 109 and 111 a pulmonary muscular artery shows the same characteristics as the rtery in the normal fetus *a* *c* In contrast to the above a characteristic muscular artery of the lung in a normal 5 mc nth old infnt shows a thin wall and a wide lumen (Fig 114a from Civin W H and Edwards J E *Ci culatio i* 2 545 1950 with permission)

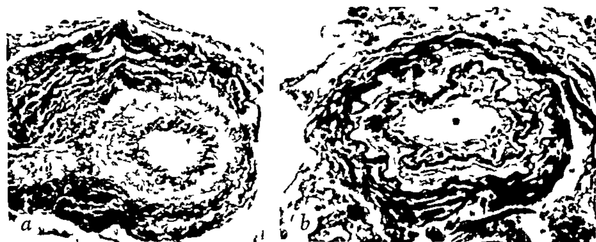


Fig 115 —Muscular artery of the l ng from the 11 month-old nfant with the Eisenmeng r complex whose specimens are illustrated n F gures 109 10) and 111 At this age the characteristic change is a th ck media a narrow lumen but a normal intima

Fig 115b—Muscular artery of the lung from Old and Russell s case of n 11 year-old patient with the Eisenmenger complex (*Am J Path* 26 789 1950 Tissues by the a thors) In contrast to the p ctu e in the younger pat ent there is superimposed int mal fibrosis The added luminal narrowing may be respons ble for the late appe ance of cy nos s n this cond t on (x375)

Relationship Between Aorta and Right Ventricle in Normal Heart

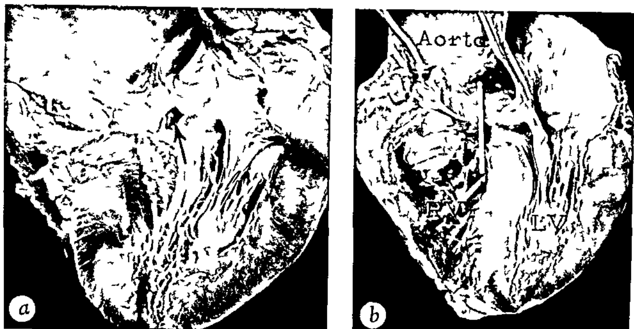


Fig 116—In this normal adult heart a membranous ventricular septal defect was artificially created at necropsy as shown in *a* (point of arrow) In *b* is shown the heart after a longitudinal section was made through the ventricular septum The probe shows the ready communication between the right ventricle (R V) and the aorta L V = left ventricle These illustrations suggest that the different functional characteristics of a case of ventricular septal defect as compared with the Eisenmenger complex depend not on a structural difference within the heart but rather on a difference in the response of the pulmonary vascular bed

Functional Studies in Venous-Arterial Shunts

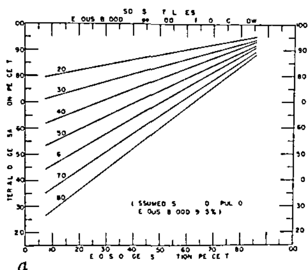


Fig 117a—A chart from which the degree of shunt or the proportions of enous and normally oxygenated blood may be read if the ear oximetric reading and a cuvette oximeter reading of the saturation of mixed venous blood have been obtained (From Burchell H B *Proc Staff Meet Mayo Clin* 5 3 7 1950 with permission)

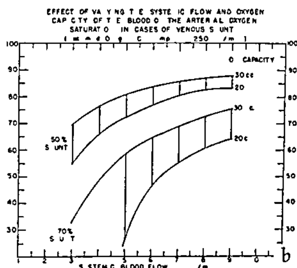


Fig 117b—Graph constructed to indicate the effect of varying the systemic flow and oxygen capacity of the blood on the arterial oxygen saturation in cases of venous arterial shunt

Stenosis of Ostium Infundibuli

STENOSIS of ostium infundibuli is characterized by a localized fibrous collar like narrowing in the outflow tract of the right ventricle The outflow tract of the right ventricle is thus divided into two portions The pulmonary valve is normal Rarely the stenosis of the infundibular ostium is the only abnormality and the condition is difficult to distinguish clinically from pulmonary valvular stenosis with intact ventricular septum In the usual case however there is an associated ventricular septal defect When this occurs the aorta may arise above the defect from both ventricles as in the case presented on the following page In such a heart the structure varies from the Eisenmenger complex only in that there is the localized zone of stenosis of the right ventricular outflow tract On this difference seems to depend the fact that the small pulmonary arteries are abnormal in the Eisenmenger complex while they were normal in the case of stenosis of ostium infundibuli presented in this report

Stenosis of Ostium Infundibuli with Biventricular Origin of the Aorta

THE PATIENT was a laborer with fairly good exercise tolerance until preterminal congestive cardiac failure at the age of 47 years An apical murmur associated with precordial thrill was present The systemic blood pressure was normal Cyanosis was not noted (From Civin W H and Edwards J E *Circulation 2* 545 1950 with permission)



Fig 118a—The left ventricle showing the ventricular septal defect above which the aorta arises



Fig 118b—The outflow tract of the right ventricle Beneath the pulmonary valve there is a localized collar like zone of stenosis which creates subpulmonary stenosis The opening of the ventricular septal defect into the right ventricle is below the zone of stenosis and is not seen in this illustration

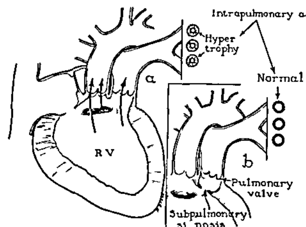


Fig 119—Comparison of the intracardiac circulation in the Eisenmenger complex (*a*) with that in stenosis of ostium infundibuli associated with biventricular origin of the aorta (*b*) The essential difference is that in the latter condition there is a localized zone of stenosis in the outflow tract of the right ventricle In the Eisenmenger complex there is no such zone of stenosis



Fig 120—A pulmonary muscular artery from the case of stenosis of ostium infundibuli with biventricular origin of the aorta illustrated in Figure 118 The artery as were all of the intrapulmonary vessels is normal Contrast this artery with those in the Eisenmenger complex illustrated in Figures 114 and 115 (Verhoeff s elastic tissue stain x130)

Tetralogy of Fallot

THE tetralogy of Fallot represents the most common malformation causing cyanosis which allows the patient to survive beyond 2 years of age The anatomic complex is characterized by biventricular origin of the aorta above a ventricular septal defect a thick right ventricle and an anatomic barrier to the flow of blood to the lungs The latter may be caused by one or more of three conditions (1) stenosis or atresia of the pulmonary trunk (2) stenosis or atresia of the pulmonary valve and (3) stenosis or atresia of the subpulmonary tract of the right ventricle The most common of these is stenosis of the subpulmonary tract of the right ventricle A right aortic arch and right descending aorta occur in about one fifth of the patients with the tetralogy of Fallot When this condition occurs the branches of the aortic arch usually are in mirror image of the normal but this is not always the case The ductus arteriosus which usually closes may lie on the right side or on the left side Rarely no ductus arteriosus is identifiable

The association of a right aortic arch with the tetralogy of Fallot is sometimes called Corvisart s disease

Tetralogy of Fallot

(Pulmonary Stenosis with Narrow Pulmonary Trunk Ventricular Septal Defect and Biventricular Origin of Aorta)

In this heart the aorta arises from both ventricles and straddles a defect of the membranous portion of the ventricular septum In contrast to the dilatation of the pulmonary trunk in the Eisenmenger complex the pulmonary trunk in this condition is narrow and there is a stenotic subpulmonic third ventricle The right ventricular wall is thick



Fig 121a—Anterior view (model x1) (1) Narrow pulmonary trunk (2) Wide aorta (3) Large right ventricle



Fig 121b—Interior of right ventricle (model x1) (1) Narrow pulmonary trunk (2) Subpulmonic stenotic third ventricle (3) Ventricular septal defect

History of the Patient

Boy 7 years old with increasing cyanosis since 5 days after birth and very limited tolerance of exercise Intense cyanosis clubbing of fingers and toes and a soft precordial systolic murmur were present Roentgenogram showed heart not enlarged with classic concavity of left border Hilar markings diminished but no hilar pulsation noted roentgenoscopically Electrocardiogram marked right axis deviation Hemoglobin 24 gm per 100 cc blood erythrocytes 8 800 000 per cu mm hematocrit 86 per cent arterial oxygen saturation 65 per cent

Principal Clinical Features of This Anomaly

1 Early cyanosis (low oxygen saturation of hemoglobin in arterial blood) 2 Paroxysmal unconsciousness with dyspnea and cyanosis a prominent feature in some cases 3 Usually a systolic murmur varying markedly in intensity from case to case 4 Clubbing of fingers and toes and polycythemia 5 Subnormal pulmonary arterial flow 6 Abscess of brain or cerebrovascular thrombosis occasionally the cause of death 7 Roentgenologic aspects usually heart is not enlarged shadow of pulmonary trunk is absent right ventricle is prominent and hilar pulsations are absent Right aortic arch not infrequently found 8 Electrocardiogram right ventricular hypertrophy



Fig 122a—Interior of the right ventricle Specimen from which models shown in Figures 121a and *b* were prepared

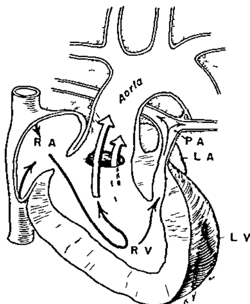


Fig 122b—Diagram of intracardiac circulation in tetralogy of Fallot



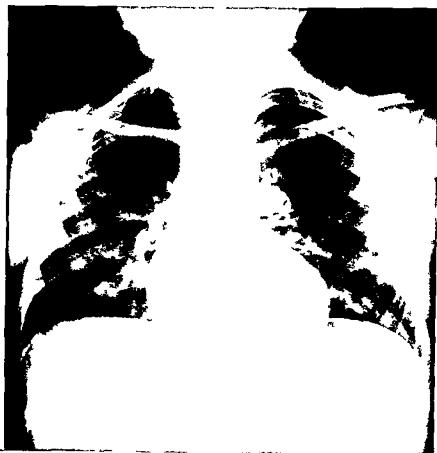


Fig 123—Electrocardiogram and thoracic roentgenogram of the patient whose heart is illustrated in Figures 121 and 122a

Tetralogy of Fallot

(Pulmonary Stenosis with Relatively Wide Pulmonary Trunk, Ventricular Septal Defect and Biventricular Origin of Aorta)

THIS HEART is also an example of the tetralogy of Fallot in spite of the relatively wide pulmonary trunk since there is stenosis at the level of the bicuspid pulmonary valve. A so-called third ventricle is also present which contributes to pulmonary stenosis.



Fig 124a—Anterior view (model x1). (1) Pulmonary trunk. (2) Large right ventricle.



Fig 124b—Interior view of right ventricle and pulmonary trunk (model x1). (1) Stenotic bicuspid pulmonary valve. (2) Subpulmonic third ventricle. (3) Ventricular septal defect.

History of the Patient

BOY 5 years old of normal development with progressive cyanosis since 3 years of age. Clubbing and mild polycythemia as well as a loud precordial systolic murmur with thrill were noted. Roentgenogram showed heart of normal size and contour but decreased hilar markings. Electrocardiogram showed right axis deviation. The boy died of abscess of the brain.

Comparison of Cases

Although the two examples of the tetralogy of Fallot shown on this and the preceding two pages differ structurally and roentgenographically in regard to the size of the pulmonary arteries, they are nevertheless identical functionally. The caliber of the pulmonary trunk may vary from moderate narrowing to atresia. When valvular stenosis is present, the pulmonary artery and the outflow tract may approach those of a normal heart. Variation is found in the length of the subpulmonic fibromuscular channel, sometimes called the third ventricle. This channel may be narrow throughout its length or it may exhibit stricture-like contractions at either end (see page 82). Enlarged bronchial arteries may carry a substantial amount of blood to the lungs in this malformation.



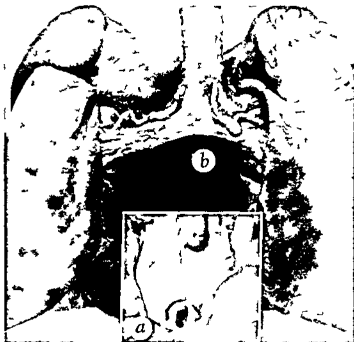
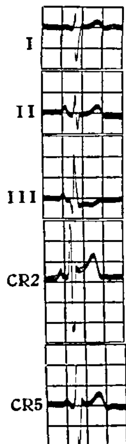


Fig 125—Inter or of the right entricle Specim n as in Figure 124

Fig 126—*a* A stenot c b cuspid pulmonary valve in a case of tetralogy of Fallot *b* Enlarged bronchial arteries in a case of tetralogy of Fallot



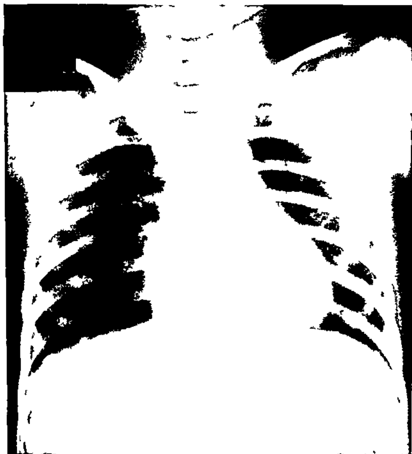


Fig 127—Electrocard ogram and roentgenogram of the patient whose heart s shown n Figures 124 and 125

Tetralogy of Fallot, Variations in Anatomic Types of Pulmonary Stenosis

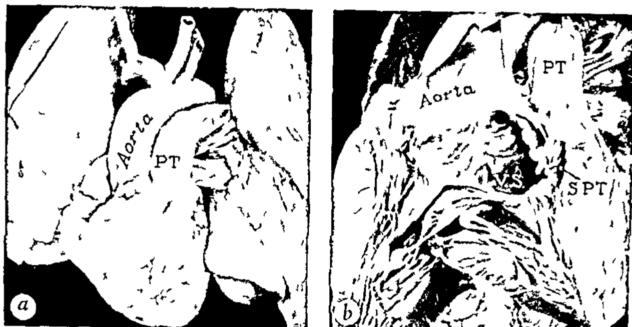


Fig 1 8—Tetralogy of Fallot in a 13 year-old boy Catheterization data are shown in Figure 13 f *a* The heart and lungs from the front The pulmonary trunk (P T) is relatively wide for this vessel in the tetralogy of Fallot The barrier to pulmonary flow is in the subpulmonary region of the right ventricle (see *b*) *b* The right ventricle Beneath the pulmonary valve there is a narrow muscular walled tract (S P T) which constitutes the basis for the barrier to pulmonary blood flow V S = ventricular septal defect P T = pulmonary trunk

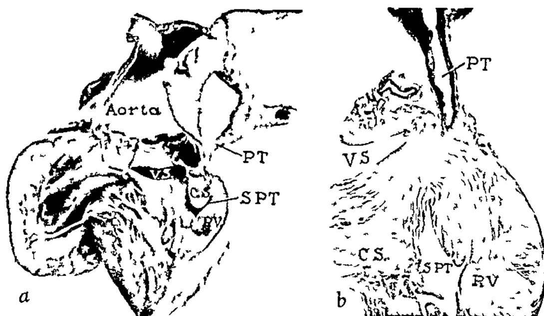


Fig 129—Tetralogy of Fallot with pulmonary atresia in a female infant 11 weeks old (Specimen submitted by Dr F Parker Jr) *a* The interior of the right ventricle showing the bi ventricular origin of the aorta above a ventricular septal defect (V S) Between the anterior wall of the right ventricle (R V) and a mass of muscle (C.S) there is the narrow subpulmonary tract (S P T) P T = pulmonary trunk *b* Photomicrograph of a section through the pulmonary trunk and subpulmonary region of the heart illustrated in *a* *The lower portion of the pulmonary trunk (P T) is atretic and does not communicate with the right* ventricle (Abbreviations as in *a*) No valvular tissue was identified in serial sections of this preparation Atresia of upper portion of the subpulmonary tract (Verhoeff s elastic tissue stain counterstained with van Gieson s connective tissue stain x4)

Tetralogy of Fallot, Pulmonary Atresia, Right Aortic Arch

CYANOTIC female infant Death at 22 days

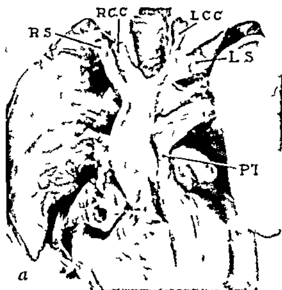




Fig 130a—(above)—Anterior view of thoracic organs Narrow pulmonary trunk (P T) above atretic valve Right aortic arch Branches mirror image of normal Left subclavian (L S) and left common carotid (L C C) arteries arise from left innominate artery R C C = right common carotid artery R S = right subclavian artery *b* Roentgenogram of thorax

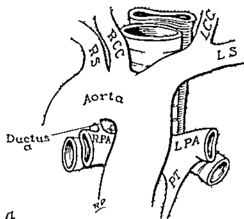
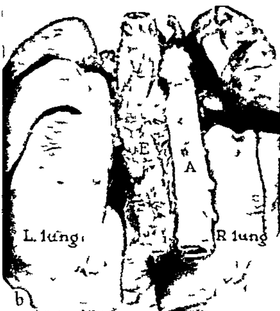


Fig 131a—(right)—Tetralogy of Fallot Right aortic arch

Fig 131b and c—(below)—*b* Thoracic organs of the case illustrated in Figure 130 from behind Descending aorta (A) lies to right of esophagus (E) *c* Right anterior view of thoracic organs showing right sided ductus arteriosus (D A) extending from right pulmonary artery (R P A) to right aortic arch R S = right subclavian artery R C C = right common carotid artery L C C = left common carotid artery L S = left subclavian artery A = ascending aorta





Tetralogy of Fallot, Right Aortic Arch

THE PATIENT was a cyanotic young woman of 18 years with the clinical and laboratory characteristics of the tetralogy of Fallot

Fig 132—The roentgenogram of the thorax shows the shadow of the right arch and the upper part of the descending aorta to the right of the midline The esophagram shows the indentation caused by the right aortic arch on the right side of the esophagus

Tetralogy of Fallot, Dye-Dilution Curves

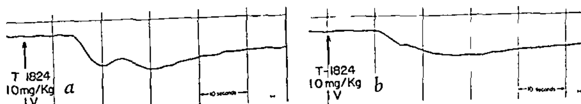


Fig 133—Dye dilution curves in a patient with the tetralogy of Fallot *a* Before operation *b* Following a Blalock Taussig anastomosis (See page 47)

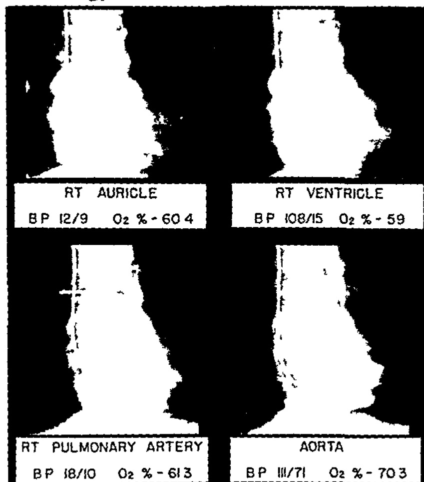


Fig 134—From the 13 year old boy with tetralogy of Fallot whose heart is illustrated in Figure 128 and functional studies in Figure 137 The position of the catheter in various locations is shown as well as the blood pressure and per cent of saturation of the blood with oxygen in the varying positions It is evident that the illustration showing the catheter in the descending aorta indicates that the catheter had passed from the right ventricle into the aorta



Fig 135—From a patient with the tetralogy of Fallot The tip of the catheter is in the aorta *a* Anteroposterior view *b* Left anterior oblique view

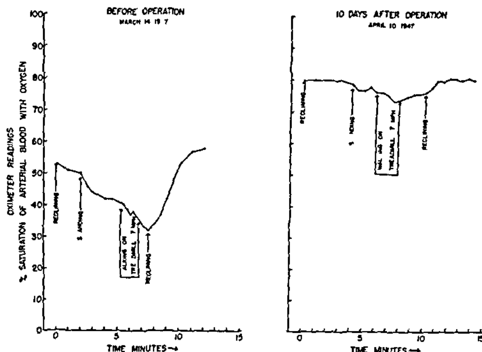


Fig 136—The effect of the Blalock Taussig anastomosis on the arterial oxygen saturation at rest and during exercise in a patient 3½ years of age with the tetralogy of Fallot Ten days after the operation the resting arterial oxygen saturation was increased 28 percentage points and the magnitude of the decrease of the arterial oxygen saturation produced by exercise was reduced from 0 to 7 percentage points (From Montgomery G E Jr *et al* *Am Heart J* 36 668 1948 with permission)

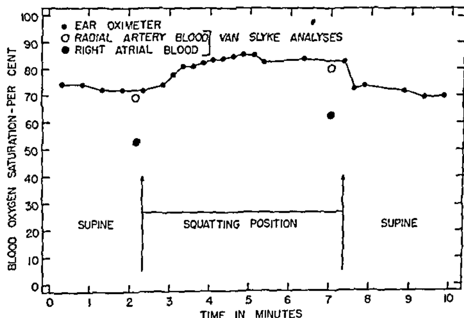


Fig 137—Effect of squatting on the arterial oxygen saturation in a 13 year old boy with the tetralogy of Fallot The gross specimens of the heart of the patient are illustrated in Figure 128 the catheterization data in Figure 134 (From Burchell H B *Proc Staff Meet Mayo Clin* 25 377 1950 with permission)

Pulmonary Stenosis with Intact Ventricular Septum

PULMONARY stenosis with intact ventricular septum is characterized by the pulmonary valves being deformed in such a way that it has the structure of a truncated cone The lumen of the valve is stenotic while the pulmonary trunk is either of normal width or dilated As the name implies the ventricular septum is intact but in somewhat more than half of the cases the atrial septum has a defect This defect takes the form either of probe patency of the foramen ovale or of a true atrial septal defect usually the former When the atrial septum is closed no shunt is possible When the atrial septum has an opening a venous arterial shunt may occur Marked right ventricular hypertrophy of the concentric type is caused by the pulmonary stenosis

Cardiac failure abscess of the brain and bacterial endocarditis are the major complications Survival to adult life is common

Pulmonary Stenosis with Intact Ventricular Septum

THE LEAFLETS of the pulmonary valve are fused to form a structure with the shape of a truncated cone Although the pulmonary trunk is wide the orifice at the level of the fused leaflets is markedly stenotic There is marked right ventricular hypertrophy In this heart there is an atrial septal defect on the basis of a probe patent foramen ovale

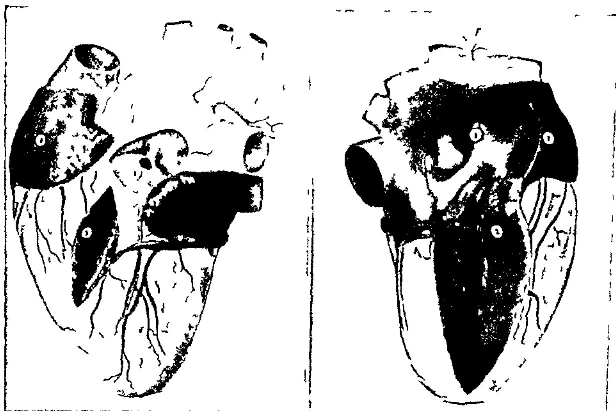


Fig 138—Model of heart ×3/4 *(a left)* Anterior view (1) Fused pulmonary leaflets causing pulmonary stenosis (2) Hypertrophied right ventricular wall (3) Right atrium *(b right)* Interior of right atrium and right ventricle (1) Edge of patent foramen ovale (2) Hypertrophied right ventricular wall (3) Right atrium

History of the Patient

MAN 26 years old left hemiplegia following severe convulsive seizures progressive increase in intracranial pressure death two weeks after craniotomy for cerebral abscess Periodic cyanosis noted at 3 years of age persistent cyanosis with clubbing since 16 years of age yet physical capacity remained only slightly sub normal Loud coarse systolic murmur with thrill third and fourth left interspace Hemoglobin 24 8 gm RBC 7 850 000 hematocrit 80 per cent x ray moderate right ventricular enlargement with prominent shadows of pulmonary artery which pulsated feebly on roentgenoscopy electrocardiogram revealed marked right ventricular hypertrophy pattern

Principal Clinical Features of This Anomaly

1 Delayed onset of cyanosis cyanosis absent if atrial septum intact dyspnea and easy fatigue with ex ertion
2 Loud coarse systolic murmur with thrill maxi mal left third interspace x ray shows enlarged right ventricle with prominent shadows of pulmo nary trunk minimal or absent pulsation on roent genoscopy
3 Electrocardiogram shows right axis deviation plus pattern of right ventricular hypertrophy in pre cordial leads
4 Arterial oxygen saturation lowered or normal
5 Catheterization very high right ventricular pres sure (may exceed systemic systolic pressure) low or normal pressure in pulmonary trunk
6 Frequent survival to adult life

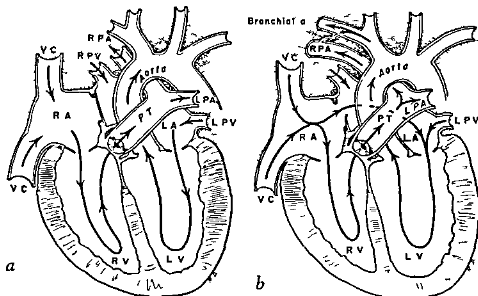


Fig 139—The intracardiac circulation in pulmonary stenosis with intact ventricular septum *a* when the atrial septum is intact there is no opportunity for a shunt *b* when there is an opening in the atrial septum a venous arterial shunt may exist Bronchial arteries may be dilated and carry blood from the aorta to the lungs (From Edwards J E *Arch Surg* 61 1103 1950 with permission)

I
II
III
CR2
CR5

Fig 140—Electrocardiogram and roentgenogram of thorax from the patient whose heart is illustrated in Figures 138 141 and 142 The prominence of the pulmonary arterial shadow is at times even greater than shown in this patient (From Parker R L M *Clin North America* 32 855 1948 with permission)

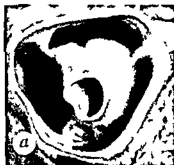




Fig 141a—(above)—The pulmonary valve from above although the pulmonary trunk is of wide diameter the cusps of the valve are fused to cause severe stenosis at valve level *b (right)* the outflow portion of the right ventricle there is pronounced concentric hypertrophy of the myocardium Perhaps as a result of this phenomenon the tract immediately inferior to the valve (containing probe) is narrow It is possible that this change may remain a barrier to the emptying of the right ventricle after an adequate valvulotomy (specimen from which models shown in Figure 138 were prepared) (Fig 141a from Parker R L M *Clin North America* 32 855 1948 with permission Fig 141b from Edwards J E *Arch Surg* 61 1103 1950 with permission)

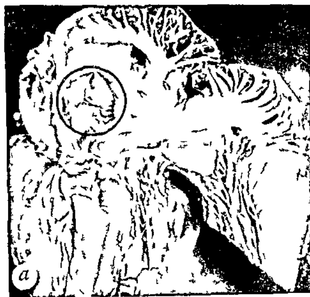




Fig 142a—(from the patient whose heart is modeled in Figure 138)—The right side of the heart the right ventricular and *atrial hypertrophy and the patent foramen ovale are shown* *b* arising from the descending aorta (A) and extending to the lungs are dilated and tortuous bronchial arteries (Br art) E Esophagus (Fig 142a from Parker R L *M Clin North America* 32 855 1948 with permission)

Pulmonary Stenosis with Intact Ventricular Septum, Clinical Case

FEMALE 19 years old Early history of progressive cyanosis and clubbing easy fatigability all symptoms accentuated past five years Harsh systolic murmur entire thorax thrill over pulmonic area Pulmonic valvulotomy performed by Dr J W Kirklin with improvement

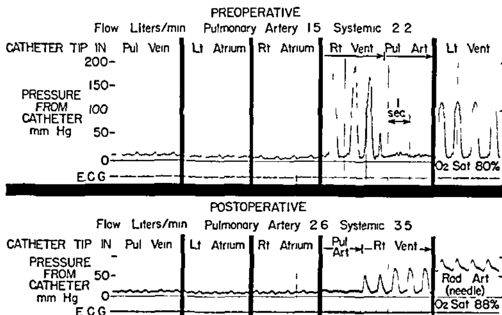


Fig 143—Cardiac catheterization studies Preoperatively the pronounced right ventricular hypertension contrasted with normal pulmonary arterial pressure Postoperatively the right ventricular pressure is significantly lower than before pulmonary valvulotomy but still above normal

Fig 144 —The preoperative electrocardiogram and roentgenogram of the thorax

Pulmonary Stenosis with Intact Ventricular Septum, Complications

AMONG the 3 complications of this malformation are cardiac failure bacterial endocarditis and abscess of the brain The last seen only in patients with an atrial septal defect or a patent foramen ovale



Fig 145—Solitary abscess of brain from man whose heart is shown on pages 88 and 91 No intracardiac infection was present (Parker R L M *Clin North America 32* 855 1948 with permission)



Fig 146—Photomicrograph of pulmonary valve with bacterial endocarditis From a 53 year-old man with valvular pulmonary stenosis (x7 5)

Pulmonary Stenosis with Relatively Minor Degree of Pressure Gradient Between Right Ventricle and Pulmonary Artery

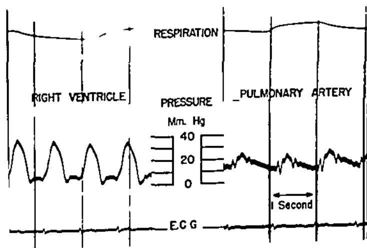


Fig 147—Pressure tracings from the right ventricle and pulmonary trunk showing evidence of mild pul (From Burchell H B and Wood E H *Proc Staff Meet Mayo Clin* 25 41 1950)

Pulmonary Atresia with Intact Ventricular Septum

(Functional Two chambered Heart)

IN PULMONARY atresia with intact ventricular septum the atresia is at valve level The leaflets of the pulmonary valves are fused to form a fibrous diaphragm like membrane The right ventricular chamber is usually small and the right ventricular wall is thick out of all proportion to the size of the chamber The tricuspid valve is small but seems *usually to function adequately An opening in the atrial septum is the* route by which venous blood which enters the right atrium escapes into the left atrium A ductus arteriosus supplies the lungs with blood This relatively uncommon malformation rarely if ever allows the patient to live beyond early infancy

Pulmonary Atresia with Intact Ventricular Septum

(*Functional Two chambered Heart*)

In this heart the pulmonary leaflets are fused to form an imperforate membrane The tricuspid valve is small but properly formed and probably functioned normally As in the case illustrated on this page the right ventricular cavity is diminutive though the right ventricular wall is tremendously thickened The only outlet for the blood entering the right side of the heart is a patent foramen ovale

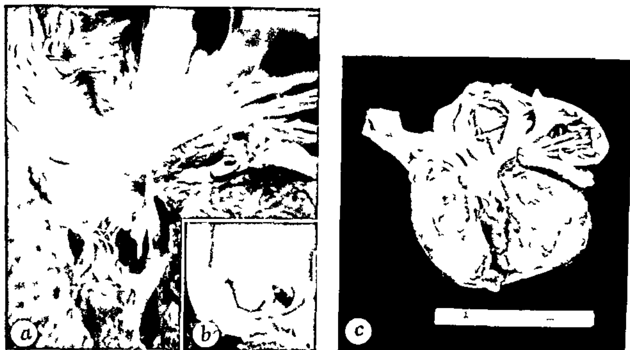


Fig 148—The right side of the heart and the pulmonary valve *a* The tricuspid valve and right ventricle The leaflets are properly formed though the valve is small *b* The pulmonary valve from above There is atresia at valve level on the basis of fusion of the three cusps to form an imperforate membrane *c* The right side of the heart Though the chamber of the right ventricle is diminutive the wall is tremendously thickened A probe lies in the patent foramen ovale the only channel by which blood can escape from the right side of the heart

History of the Patient

Male 11 months old cyanotic since birth retarded development and marked malnutrition Soft systolic murmur at left sternal border Roentgenogram enlarged boot shaped heart absent shadow of pulmonary trunk clear lung fields Electrocardiogram infantile type with large biphasic QRS complexes Hemoglobin 17 3 gm RBC 6 000 000 Died suddenly following syncopal attack associated with dyspnea

Principal Clinical Features of This Anomaly

1 Cyanosis at birth with progressive increase in severity associated with dyspnea
2 Progressive cardiac enlargement X ray absent shadow of pulmonary trunk clear lung fields aorta appears enlarged Configuration similar to tetralogy of Fallot except for enlargement of heart
3 Systolic murmur not definitive in type
4 Electrocardiogram large biphasic QRS complexes right axis deviation
5 Death in early infancy survival seldom more than a few months

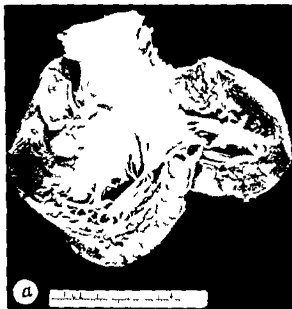
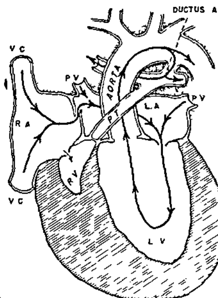


Fig 149a—The left ventricle The aorta is in free communication with the enlarged left ventricle Th s chamber repre ents funct onally a single chamb r



F 149b The intracardiac c rculation n pulmonary atresia with closed entricular septum Blood flow to the lung depends pr marily on a patent duct s arteriosus

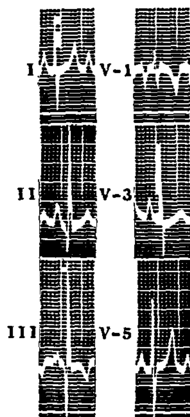




Fig 150—The electrocardiogram a d the roentgenog am of the thorax from the patient whose heart is illustrated in Figures 148 and 149

Pulmonary Atresia with Intact Ventricular Septum and Enlarged Right Ventricular Chamber

JUDGING from the case illustrated and described on pages 94 and 95 and from the few reported examples of pulmonary atresia with intact ventricular septum there is usually a small right ventricular chamber with a thick right ventricular wall The case illustrated in Figure 151 differs from this description in that the right ventricular chamber is wide as is the tricuspid orifice The right atrium is also greatly dilated



Fig 151—Right side of heart in a case of pulmonary atresia with intact ventricular septum showing dilatation of the right ventricular chamber and of the tricuspid orifice The right atrium is markedly dilated (Specimen contributed by Dr Ted E Ludden)

Complete Transposition of the Great Vessels

COMPLETE transposition of the great vessels is characterized by origin of the aorta from the right ventricle and of the pulmonary trunk from the left ventricle The two great vessels lie parallel to each other the aorta in front of the pulmonary trunk This creates a narrow vascular shadow in the roentgenogram when the great vessels are viewed from the front

The venous connections of the heart are normal therefore the abnormal arterial connections lead to a profound circulatory disturbance in that venous blood does not have a ready route to the lungs Unless there is some communication between the two circulations life after the umbilical cord is divided is not possible The usual communications are in the form of a patent foramen ovale a ventricular septal defect a patent ductus arteriosus or more than one of these The bronchial arteries may also serve as a route by which venous blood is carried to the lungs

This malformation has a tendency to occur in the male sex Survival beyond infancy is rare

Complete Transposition of the Great Vessels

In this heart the aorta arises from the right ventricle and the pulmonary artery from the left ventricle A defect of the membranous portion of the ventricular septum is present in this specimen The foramen ovale and the ductus arteriosus are patent



Fig 15 a—Anterior view (model x1) (1) Aorta situated anterior to (2) pulmonary trunk



Fig 15 b—Left lateral view (model x1) (1) Ventricular septal defect (2) Aorta arising from right ventricle (3) Pulmonary trunk arising from left ventricle

History of the Patient

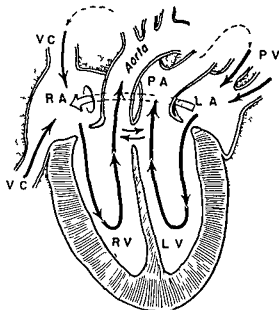
A Female 2 months old had had intense cyanosis at birth which disappeared Intermittent cyanosis recurred at 5 weeks The patient was hospitalized for pneumonia at 2 months Cyanosis and dyspnea increased with evidence of congestive heart failure A systolic murmur over the precordium and interscapular area was noted Roentgenogram of the thorax showed globular cardiac enlargement with abnormally narrow shadows of the great vessels in the A P view Electrocardiogram showed high voltage biphasic QRS complexes Progressive respiratory embarrassment and death ensued

Principal Clinical Features of This Anomaly

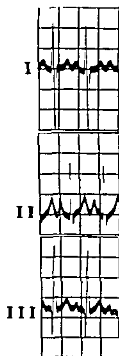
1 Cyanosis onset at birth severe and progressive
2 Usually death in early infancy
3 Progressive cardiac enlargement
4 Variable systolic murmur
5 Roentgenologic aspects usually right ventricular enlargement globular configuration aorta anterior to pulmonary trunk Shadow of great vessels narrow in A P view
6 Electrocardiogram usually right axis deviation occasionally left axis deviation



Fig 153—Specimen from which models shown in Figure 152 were prepared Interior of the right ventr cle and aorta



F g 154—Diagram of intracard ac circulation in com plete transposit on of great vessels with ventricular septal defect





F g 155—El t card gram nd th racic ro ntge ogram of the patient whose he rt s llustrated in F g es 15 and 153

Complete Transposition of the Great Vessels
Case with Unusual Longevity

THE AORTA arises from the right ventricle The pulmonary trunk arises from the left ventricle The coronary arteries arise from the aorta There is a ventricular septal defect The ventricular walls are hypertrophied



Fig 156a—Right ventricle and aorta arising from this chamber



Fig 156b—Left ventricle showing origin of pulmonary trunk from this chamber Ventricular septal defect

History of the Patient

GIRL 7½ years old Normal birth and development until 15 months of age when cyanosis and dyspnea became evident on exertion Progressive persistent cyanosis and clubbing since 3 years of age associated with poor exercise tolerance

Examination at Five Years

Harsh systolic murmur maximal left second interspace Hematocrit 80 per cent Arterial oxygen saturation 70 per cent Chest x ray revealed cardiac enlargement with marked prominence of vascular hilar shadows Electrocardiogram revealed marked right ventricular hypertrophy Exploration revealed high pulmonary arterial pressure no anastomosis attempted Patient died from injuries sustained in auto accident 2 years later at age of 7½ years

Synopsis of Significant Catheterization Data*

	Pressure mm Hg	O_2 saturation per cent
Superior vena cava	18/7	55.5
Inferior vena cava	10/1	53.5
Atrium (left ?)	12/7	87.0
Atrium (right)	18/5	57.5
Ventricle	96/7	6[illegible]
Aorta innominate artery	89/56	64.0
Radial artery	84/58	64.0

* Under rectal avertin and $N_2O + O_2$ anesthesia Continuous determination of *arterial saturation by ear oximeter* was not a routine practice when this patient was studied (April 1948)



Fig 157a—The left side of the heart There is a large atrial septal defect (in circle) The coronary sinus (C S) has been divided It is wide on the basis of having received a persistent left superior vena cava which was an additional and incidental malformation



Fig 157b—Exterior of heart viewed from left The aortic origin (A) lies in front of that of the pulmonary trunk (P T) Both auricular appendages (R A = right auricular appendage L A = left auricular appendage) lie to the left of the great arterial vessels The cut lower end of the persistent left superior vena cava (L V C) just before its entrance into the coronary sinus is shown

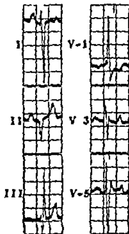


Fig 158—The electrocardiogram



Fig 159a—The cardiac catheter has passed through the right side of the heart into the aorta and then into the innominate artery

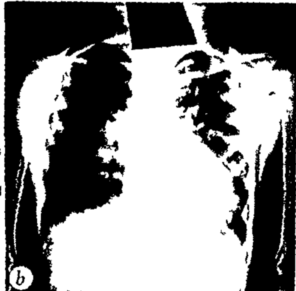


Fig 159b—The roentgenogram of the thorax All illustrations on this page are from case described on preceding page



Fig 160—From a cyanotic 1 month old male infant *a* External view of heart and great vessels from the front The left auricular appendage (L A) forms a prominence along the left cardiac margin Superior vena cava (S V C) to right of ascending aorta (A) *b* The roentgenogram of the thorax The shelflike prominence of the left cardiac margin seems to be caused by the shadow of the left auricular appendage Narrow shadow of great vessels

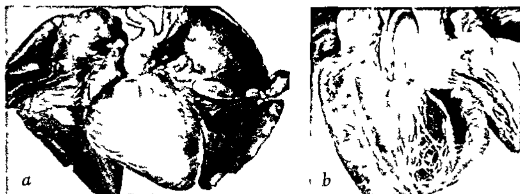


Fig 161—From a cyanotic male infant 1 month old *a* Anterior view of heart and lungs Transposed aorta obscures pulmonary trunk which lies behind it *b* Origin of pulmonary trunk from left ventricle

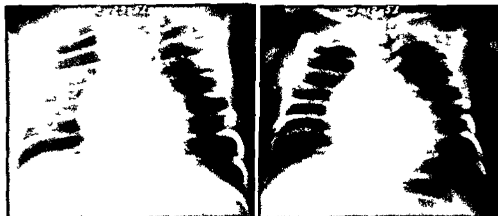


Fig 16 —From the patient whose heart is illustrated in Figure 161 showing evidence of rapidly progressing cardiac enlargement

Corrected Transposition of the Great Vessels

CORRECTED transposition of the great vessels as the name implies is an anomaly in which the aorta communicates with the left ventricle and the pulmonary trunk communicates with the right ventricle but the aorta is anterior to the pulmonary trunk as it is in complete transposition of the great vessels The transposition may thus be considered as cor rected since there is no functional disturbance in spite of this gross anatomic abnormality A ventricular septal defect is usually present in corrected transposition of the great vessels Under unusual circum stances the ventricular septum may be intact and when such is the case the patient has no cardiac disability The case portrayed in the next three pages is an example of such a phenomenon Mitral in sufficiency from an incompetent left A V valve may be observed in some cases

Corrected Transposition of the Great Vessels

In the normal heart the pulmonary trunk arises from the right ventricle anterior to the aorta The two vessels rotate about each other In corrected transposition while the great arteries connect with the appropriate ventricles the aorta arises anterior to the pulmonary trunk and the vessels fail to spiral about each other The venous connections are normal The patient with corrected transposition of the great vessels whose heart is illustrated on this page and on pages 105 and 106 was a man 47 years of age who died of intestinal obstruction No symptoms of cardiac disease had been present The specimen of the heart was contributed by Dr W B Chamberlin Jr For a similar case but associated with a patent ductus arteriosus see page 118

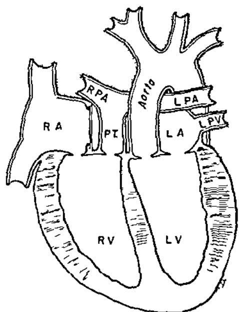


Fig 163—Diagrammatic representation of heart and great vessels in corrected transposition of the great vessels

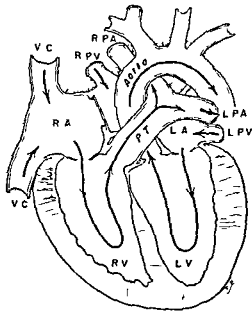


Fig 164—Diagrammatic representation of normal heart and great vessels

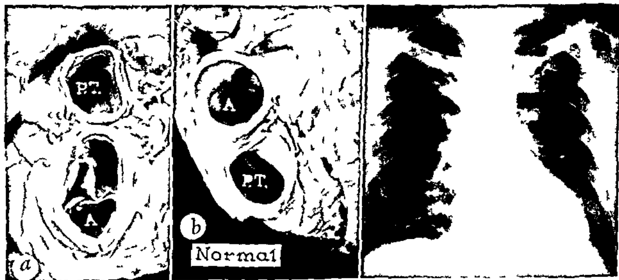


Fig 165a—(left)—The aorta and pulmonary trunk in corrected transposition of the great vessels The aorta (A) lies anterior to the pulmonary trunk (P T) *b (center)*—The aorta and pulmonary trunk in a normal heart The pulmonary trunk (P T) lies anterior to the aorta (A)

Fig 166—(right)—Roentgenogram of the thorax in the patient with corrected transposition of the great vessels (Roentgenogram contributed by Dr J V Pischieri)



Fig 167a—The right side of the heart in corrected transposition of the great vessel The foramen ovale is normally formed The right A V valve resembles a normal mitral valve (see Fig 168*b*) and is dissimilar to a normal tricuspid valve illustrated in *b*

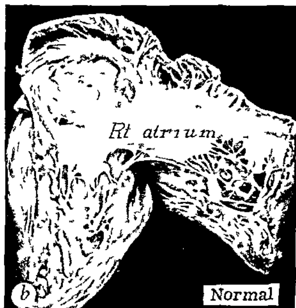


Fig 167b—Right side of heart and tricuspid valve of a normal heart See Figures 169 and 170 for illustrations of ventricles and great vessels



Fig 168a—Left side of heart in corrected transposition of the great vessels The atrial septum is normal The left A V valve does not resemble a normal mitral valve as illustrated in *b* and is similar in appearance to a normal tricuspid valve (see Fig 167*b*)



Fig 168b—The left side of heart and mitral valve from a normal heart



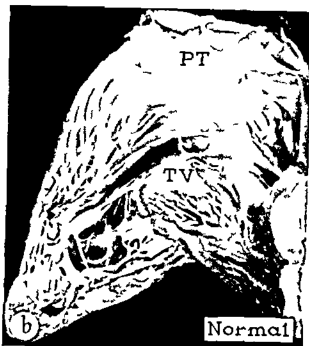


Fig 169a—Right sided ventricle in corrected transposition of great vessels The pulmonary valve is intimately related to the right A V valve *the relationship is similar to the relationship of the aortic valve to the mitral valve in the* normal heart (see Fig 170*b*)

Fig 169b—Right ventricle and tricuspid and pulmonary valves of a normal heart PT = pulmonary trunk TV = tricuspid valve



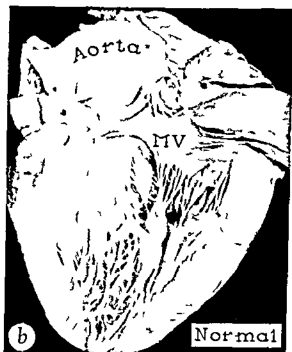


Fig 170 —Left sided ventricular chamber and aortic valve in *corrected transposition of the great* vessels *The* left *A* V valve and the aortic valve fail to have the intimate association seen in normal hearts as illustrated in *b* The relationship between these two valves resembles that between the pulmonary and tricuspid valves of normal hearts (see Fig 169*b*) The trabeculated appearance of the left sided ventricle is similar to that seen in a normal right ventricle and dissimilar to the appearance in a normal left ventricle No ventricular septal defect

Fig 170b—The left ventricle and mitral (M V) and aortic valves of a *normal heart*

Isolated Dextrocardia

WHEN DEXTROCARDIA is associated with complete situs inversus or the mirror picture of normal organ arrangement the heart is usually functionally normal and anatomically a mirror image of the normal It is generally recognized that dextrocardia without an associated situs inversus usually involves serious intracardiac malformation Likewise the rare isolated levocardia with an otherwise complete situs inversus is often associated with intracardiac defects The right sided hearts with serious malfunction without total situs inversus characteristically how ever have a situs inversus arrangement of the atria and ventricles There exist cases of apparent dextrocardia without associated situs inversus in which the heart from the functional viewpoint is normal In these in stances the incongruous cardiac asymmetry in relationship to the general splanchnic arrangement usually follows a different pattern from that of the normal situs inversus heart The question indeed has arisen as to the propriety of the nomenclature and in the case chosen herein for presentation it has been suggested that dextrotorsion of the heart might be more apt than dextrocardia Another term applied to this type of heart has been dextrocardia without inversion of the cardiac cavities The basic classification of dextrocardia usually employed is that pro posed by Mandelstamm Moritz and Reinberg Samuel (*Ergebn d inn Med u Kinderh* 34 154 1928)

Isolated Dextrocardia

A BOY 15 years old always healthy Chest survey x ray heart out of place Physical examination maximal location of the first heart sound in the midsternal area (From Burchell H B and Pugh D G *Am Heart J* 44 196 1952 with permission)



Fig 171a and b—Roentgenogram of the thorax and electrocardiogram of the patient

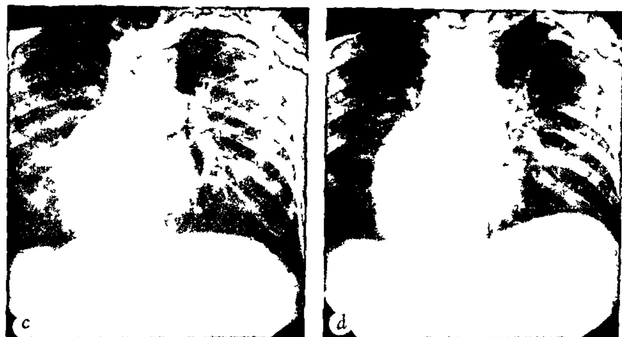


Fig 171c and d—Angiocardiograms taken by Dr D G Pugh *c* Film two seconds after injection of dye showing right atrium right ventricle and pulmonary arteries *d* Film at 6½ seconds additionally filling of left side of heart and aorta

Electrocardiogram in Situs Inversus



Fig 172—Electrocardiogram in situs inversus complexes in lead 1 inverted Precordial leads taken in conventional manner Inversion of T waves in precordial leads over the left chest and normal complexes in precordial leads over the right chest (From Dry T J *A Manual of Cardiology* Philadelphia W B Saunders Company 1943 p 161 with permission)

Persistent Truncus Arteriosus

PERSISTENT truncus arteriosus is characterized by a single functioning vessel leaving the heart and this vessel receives the blood above a ventricular septal defect from both ventricles This vessel gives origin to the coronary systemic and pulmonary circulations The pulmonary arteries may arise independently and directly from the truncus arteriosus or from a pulmonary trunk In one type no pulmonary arteries as such exist the lung being supplied by bronchial arteries

True persistent truncus arteriosus should be distinguished from cases of atresia of the pulmonary trunk wherein the pulmonary trunk is either absent or represented by an atretic cord The right and left pulmonary arteries in such cases receive blood from the aorta through a patent ductus arteriosus as in case presented on page 95 and in Figure 177 page 112

Aorticopulmonary septal defect is a variation of persistent truncus arteriosus in which only a small communication exists between the ascending aorta and the pulmonary trunk (pages 112 and 117)

Persistent Truncus Arteriosus

In this heart a single arterial vessel the persistent truncus arteriosus arises from both ventricles above a ventricular septal defect An incomplete septum divides the upper portion of the truncus arteriosus into the aorta and a short pulmonary trunk No vestige of the ductus arteriosus is present in this specimen



Fig 173a—Anterior view (model x1) (1) Persistent truncus arteriosus (2) Large right ventricle



Fig 173b—Interior of truncus arteriosus and right ventricle (model x1) (1) Incomplete truncus septum (2) Ventricular septal defect below biventricular origin of persistent truncus arteriosus

History of the Patient

Male infant 8 months old systolic murmur present since birth There was feeding difficulty with poor development The patient was admitted one day before death A loud systolic murmur was heard over the entire precordium dyspnea and swelling of the hands and feet were noted the liver was palpable but there was no cyanosis The thoracic roentgenogram showed marked cardiac enlargement with a narrow vascular shadow The electrocardiogram showed right axis deviation The infant died suddenly

Principal Clinical Features of This Anomaly

1 Cyanosis usually moderately severe but occasionally absent Oxygen saturation of arterial hemoglobin always below normal the degree of desaturation depending on pulmonary blood flow and perhaps the effect in some cases of directional ejection from the ventricles
2 Loud systolic murmur occasionally continuous murmur
3 Roentgenologic aspects right and left ventricular enlargement Increased pulsation of hilar shadows may be present particularly when cyanosis is minimal
4 Electrocardiogram usually right axis deviation



Fig 174a—Specim n from which models shown in Figure 173 were prepared Interior of right ventricle and persistent truncus arteriosus

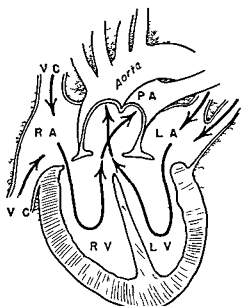


Fig 174b—Diagram of intracardiac circulation in persistent truncus arteriosus

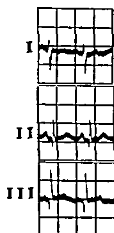




Fig 175—Electrocardiogram and thorac c roentgenogram of the patient whose heart is illustrated in Figures 173 and 174a

Persistent Truncus Arteriosus, Anatomic Classification

(From Collett R W and Edwards J E S Clin North America 29 1245 1949 with permission)

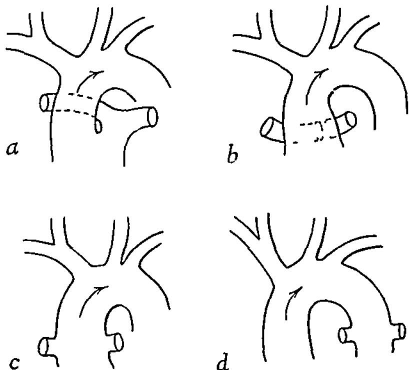


Fig 176—*a* Type 1 A single pulmonary trunk and ascending aorta arise from the truncus arteriosus
b Type 2 *The right and left pulmonary arteries arise close together from the dorsal wall of the truncus arteriosus*
c Type 3 One or both pulmonary arteries arise independently from either side of the truncus arteriosus
d Type 4 There are no pulmonary arteries and there is apparent congenital absence of the sixth aortic arches Th arterial circulation to the lungs is by way of bronchial arteries



Fig 177—Single arterial trunk with pulmonary arteries supplied through patent ductus arteriosus This condition sometimes referred to as persistent truncus arteriosus in reality *should not be so classified In short the pulmonary trunk* has entirely disappeared as a recognizable structure or at times is overlooked because it resembles a fibrous strand similar to the appearance of the pulmonary trunk illustrated in Figure 4 (page 3)



Fig 178—Partial persistent truncus arteriosus (aorticopulmonary septal defect) In this condition the aortic and pulmonary valves are normally formed A fistulous opening *exists between the ascending aorta and the pulmonary trunk* Functionally this condition resembles patent ductus arteriosus and may give clinical signs readily confused with that condition (see Fig 188 page 117)

Patent Ductus Arteriosus

NORMALLY the ductus arteriosus a channel which short circuits blood from the pulmonary artery to the aorta during fetal life becomes obliterated shortly after birth Sometimes this fails to occur giving rise to the clinical entity patent ductus arteriosus The flow of blood now is from the aorta into the pulmonary artery It may reach great volumes ranging in rare instances up to as much as three fourths of the left ventricular output A patent ductus arteriosus is easily recognized in most cases by the presence of a continuous or machinery murmur The effects of a patent ductus are usually reflected in left ventricular dilatation and hypertrophy

There is usually no material elevation of pulmonary arterial pressure because of the normally low resistance of the pulmonary vascular bed but in some patients pulmonary hypertension may develop as a result of intrapulmonary arterial changes When this occurs the electrocardiographic features of right ventricular hypertrophy may appear and the characteristic machinery murmur may disappear

Among the chief causes of death in cases of untreated patent ductus arteriosus are left ventricular failure and bacterial infection involving either the patent ductus or the left pulmonary artery opposite the mouth of the ductus The patient usually lives to adult life although life expectancy is materially reduced In an uncommon instance cardiac failure may occur during infancy The continuous murmur of patent ductus arteriosus is usually absent in infancy Evidence of progressive cardiac failure in an acyanotic infant should suggest the possibility of this condition

The closure of a patent ductus arteriosus has become a standard surgical procedure the risk of which is far less than the risk of leaving this anomaly untreated

Patent Ductus Arteriosus

(Cylindric and Window Types)

THESE two hearts illustrate two anatomic types of patent ductus arteriosus the cylindric type and the window type As a consequence of the shunt from the aorta to the pulmonary arterial system there is dilatation of the pulmonary arteries and the left side of the heart



Fig 179—Cylindric type (model x1) (1) Patent ductus arteriosus (2) Dilated right ventricular outlet



Fig 180—Window type (model x½) (1) Window type patent ductus arteriosus (2) Dilated right ventricular outlet (3) Dilated left pulmonary artery

History of These Patients

A MALE 6 weeks old (Fig 179) had had a basal systolic murmur and cardiac enlargement since birth (premature) with feeding difficulty and poor development Sudden respiratory embarrassment with terminal cyanosis occurred In a man 45 years old (Fig 180) a continuous murmur had been heard when he was 30 years old Cardiac failure recurred intermittently for three years he died suddenly of pulmonary embolism On his last admission a loud systolic apical murmur (typical ductus murmur absent) and auricular fibrillation were noted Systolic blood pressure was 110 diastolic 70 A thoracic roentgenogram showed cardiac enlargement with a prominent pulmonary trunk Electrocardiograms showed a change to right axis deviation

Principal Clinical Features of This Anomaly

1 Continuous arteriovenous fistula type of murmur usually associated with thrill maximal in left second and third interspaces
2 In infants usually only a systolic murmur
3 No cyanosis normal arterial oxygen saturation
4 Increased pulse pressure (collapsing pulse)
5 Increased oxygen content of pulmonary arterial blood relative to right ventricular blood (cardiac catheterization)
6 Roentgenologic aspects the cardiac silhouette frequently appears normal Usually there is a prominent left ventricle and pulmonary artery and increased hilar pulsations In infants with symptoms progressive cardiac enlargement
7 Electrocardiogram normal or left axis deviation in usual cases Evidence of right ventricular hypertrophy in cases complicated by pulmonary hypertension



Fig 181—Specimen from which the model shown in Figure 1 9 was prepared



Fig 182—Specimen from which model shown in Figure 180 was prepared Interior of the left pulmonary artery Mouth of ductus in circle

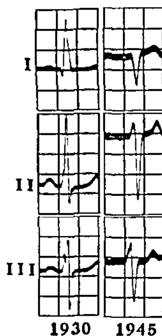




Fig 183—Electrocardiogram and thoracic roentgenogram of the patient whose heart and great vessels are shown in Figures 180 and 18

Patent Ductus Arteriosus, Clinical Case

(Chosen to Demonstrate More Typical Clinical Picture)

A MAN 38 years old Normal since childhood Good exercise tolerance Continuous murmur over pulmonary area Blood pressure 118/71 Patent ductus arteriosus demonstrated at operation Ductus successfully ligated

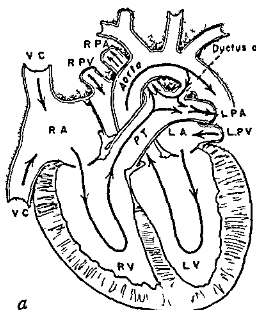


Fig 184a—Circulation in patent ductus arteriosus

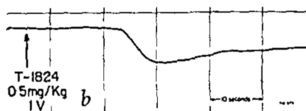


Fig 184b—The dye dilution curve of this patient (see page 47 and contrast with Fig 186a)

Synopsis of Catheterization Data

	Pressure mm Hg	O saturation per cent
Superior vena cava	7/3	69
Inferior vena cava	10/7	75
Right atrium	8/0	73
Right ventricle (outflow)	34/9	72
Pulmonary trunk	30/ 0	83
Flow liters/min /M	Systemic ? 9	Pulmonary 5 0

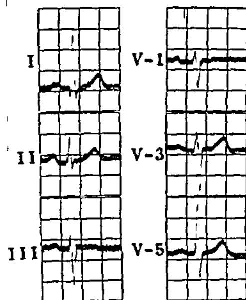


Fig 185—The electrocardiogram and the roentgenogram of the thorax of the patient whose history and functional data are summarized on this page

Patent Ductus Arteriosus with Pulmonary Hypertension

MAN 35 years old Known cardiac murmur since childhood At 30 years congestive failure Prolonged diastolic murmur over pulmonic area Functional studies revealed pulmonary hypertension and patent ductus arteriosus with reversibility of flow The ductus was ligated Condition subsequently fairly satisfactory

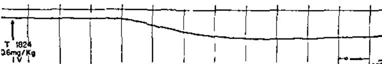
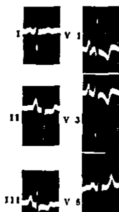
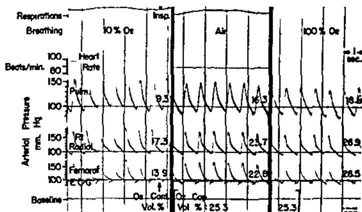




Fig 186—a (above)—The dye dilution curve Evidence of pulmonary recirculation and cardiac failure Contrast with Figure 184b *b (right)* Roentgenogram taken during cardiac catheterization Catheter has passed through the patent ductus arteriosus and has entered the descending aorta

Fig 187—(below)—(left)—The electrocardiogram *(right)* Functional studies indicating pulmonary hypertension and intermittent reversed flow





Aorticopulmonary Septal Defect





Fig 188—Roentgenogram taken during catheterization in the anteroposterior and lateral views when the catheter had passed through a communication between the pulmonary trunk and the ascending aorta Tip of catheter in ascending aorta From a 6 year old boy (Catheterization performed and illustrations submitted by Dr Forrest H Adams)

Patent Ductus Arteriosus, Corrected Transposition of the Great Vessels

GIRL 3 years old Respiratory difficulties in infancy Loud systolic and diastolic murmurs (questionably continuous) to the left of lower part of sternum and prolonged thrill No intracardiac left to right shunt (cardiac catheterization) Pulmonary trunk not entered by the catheter Pressure in right ventricle 68/3 systemic pressure 87/52 The dye curve suggested pulmonary recirculation

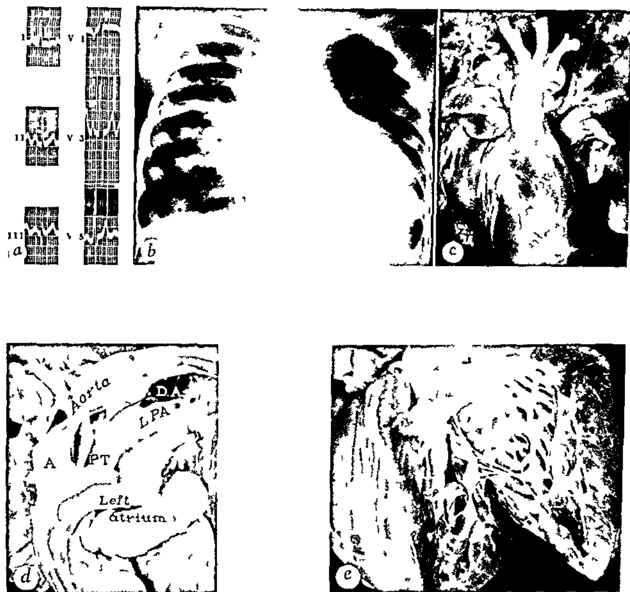


Fig 189—*a* The electrocardiogram *b* Roentgenogram of the thorax *c* The heart and great vessels from the front The aorta lies in an anterior position obscuring view *of the pulmonary trunk which lies directly behind it* *d* Lateral view of the heart and opened great vessels The ascending aorta (A) lies in front of and parallel to the pulmonary trunk (P T) A patent ductus arteriosus (D A) runs between the left pulmonary artery (L P A) and the descending aorta *e* The aorta communicates with the left sided ventricle *Its relation to the left A V valve is abnormal and is typical for that in* corrected transposition (see Figs 167 170 pages 105 106) No ventricular septal defect

Patent Ductus Arteriosus, Cardiac Failure at 8½ Months of Age

GIRL 8½ months old. History of mild blue spells during crying. Systolic murmur first noted at age of 2 months. Blue spells continued with increasing dyspnea and evidence of cardiac failure. Progressive increase in size of heart. Died of congestive heart failure.

Fig 190a—Great arterial vessels showing a patent ductus arteriosus. P T = pulmonary trunk. D A = patent ductus arteriosus. *b* The electrocardiogram.



Fig 190c—Roentgenograms of the thorax showing progressive cardiac enlargement. The first film was taken when the infant was 4 days old, the second when 2½ months old, and the third at the age of 8 months. (The first two roentgenograms submitted by Dr G S Owen.)

Patent Ductus Arteriosus, Bacterial Endarteritis

MAN 23 years old Deformity of the heart known since infancy Nov 1945 fever and loss of weight Treated with penicillin In April 1946 symptoms recurred *Typical findings of patent ductus* Treated with penicillin and then thorax explored Massive hemorrhage occurred while ductus being exposed

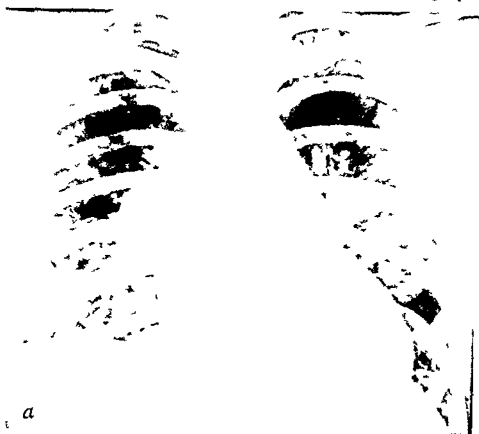


Fig 191a—Roentgenogram of the thorax showing shadows of pulmonary infarcts complicating bacterial endarteritis of patent ductus arteriosus





Fig 191b and c—Photomicrographs of lung *b* Abscesses (H + E × 6) *c* A pulmonary artery is occluded by an organized thrombus interpreted as an embolus from th inflammatory patent ductus ×15

Patent Ductus Arteriosus with Obstructive Pulmonary Vascular Lesions

WOMAN 23 years old History of dyspnea on exertion since childhood Once told she had a patent ductus arteriosus At 21 years toxemia of pregnancy Three months after birth of child congestive cardiac failure developed Responded only temporarily to treatment Harsh systolic murmur best heard at left border of sternum Died of congestive cardiac failure (From Douglas J M *et al* *Proc Staff Meet Mayo Clin* *22* 413 1947 with permission)

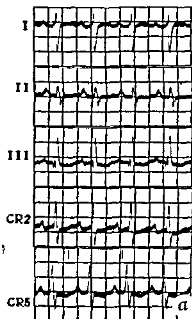




Fig 192a and b—a The electrocardiogram *b* The pulmonary artery patent ductus and aorta





Fig 192c and d—c Small elastic artery of the lung showing pronounced intimal fibrous proliferation (elastic tissue stain x75) *d* Muscular artery of the lung showing mild medial hypertrophy and pronounced intimal fibrous proliferation causing pronounced luminal narrowing (Verhoeff s elastic tissue stain counterstained with van Gieson s connective tissue stain x75)

Patent Ductus Arteriosus Complicated by Aneurysm of the Right Pulmonary Artery

A 37 year old man chronic cough Ten months ago believed to have enlarged mediastinal nodes for which roentgen therapy given No cardiac murmurs thrills or enlargement Serology negative Expansile mass in the right perihilar region calcification in walls Diagnosis aneurysm of pulmonary artery Thorax explored Size of aneurysm precluded surgical attack Death on third postoperative day (From Deterling R A Jr and Clagett O T *Am Heart J* 34 471 1947 with permission)



Fig 193a and b—Roentgenograms of the thorax showing shadow of aneurysm of right pulmonary artery Patent ductus is the commonest malformation associated with aneurysm of a pulmonary artery yet even then it is uncommmon

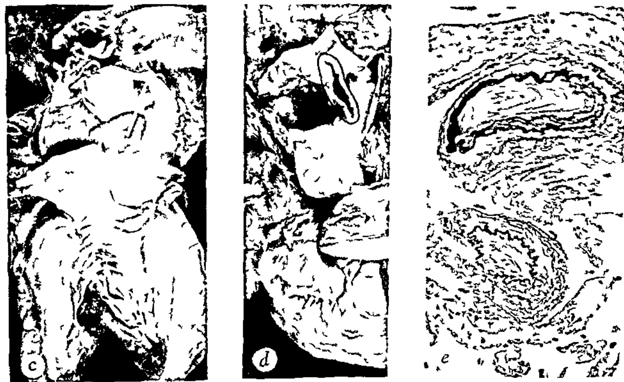


Fig 193c d and e—*c* Hypertrophied right ventricle and pulmonary trunk Probe in pulmonary ostium of patent ductus arteriosus *d* Within ring is an unruptured tear in the wall of the aneurysm of the right pulmonary artery *e* Photomicrograph of pulmonary arteries showing pronounced intimal fibrous thickening with associated luminal narrowing (x170)

Aneurysm of Aortic Sinus

(Aneurysm of Sinus of Valsalva)

ANEURYSM of an aortic sinus is a rare condition most commonly involving the right aortic sinus and less commonly the posterior aortic sinus It is doubtful whether congenital aneurysms of the left aortic sinus ever occur The aneurysm presents toward the right atrium or the right ventricle It may communicate congenitally with either of these chambers or there may be no communication on a developmental basis Acquired communication between the aorta on one hand and the right atrium or right ventricle on the other may appear spontaneously or as a complication of bacterial infection of the aneurysm

The functional disturbance from an aneurysm of an aortic sinus which communicates with one of the right sided cardiac chambers is comparable to that from patent ductus arteriosus and peripheral signs suggesting aortic insufficiency may be prominent

Aneurysm of the Right Aortic Sinus Communicating with the Right Ventricle, Ventricular Septal Defect

IN THIS heart is an aneurysm of the right aortic sinus communicating with the right ventricle A ventricular septal defect is also present

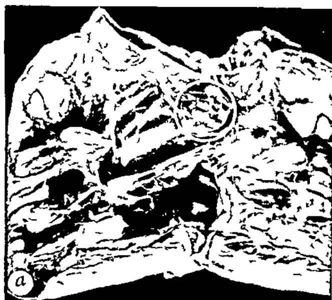


Fig 194a—The right ventricle Beneath the pulmonary valve an aneurysm of the right aortic sinus (within circle) The right ventricle hypertrophied and dilated



Fig 194b—The aneurysm of the right aortic sinus seen in *a* is shown in greater detail Several perforations in the aneurysm Beside aneurysm is a crescent shaped ventricular septal defect (V S D) (Specimen submitted by Dr Fred Sloan Clinical aspects of case reported by Morgan E H and Burchell H B *Proc Staff Meet Mayo Clin* 25 69 1950 and pathologic aspects by Burchell H B and Edwards J E *Proc Staff Meet Mayo Clin* 26 336 1951 with permission)

History of the Patient

MAN 34 years old heart murmur discovered at 5 years Impaired physical endurance and dyspnea with exertion since childhood No cyanosis Examination Systolic thrill and loud continuous murmur maximal in left second interspace transmitted widely Blood pressure 180/60 0 Pistol shot wounds over femoral arteries Roentgenogram cardiac enlargement marked prominence and increased pulsation of pulmonary arterial shadows Electrocardiogram sinus rhythm left ventricular hypertrophy Died of congestive heart failure

Principal Clinical Features of This Anomaly

1 If aneurysm does not communicate with a cardiac chamber no symptoms or abnormal signs are present
2 If aneurysm communicates with a right sided cardiac chamber from birth there may be a continuous murmur pulmonary congestion and prominent pulmonary vascular markings
3 If aneurysm ruptures postnatally usually sudden onset of severe dyspnea a continuous murmur collapsible pulse and high pulse pressure in systemic arteries Roentgenogram shows prominent pulmonary arterial shadows and pulmonary congestion Death early after postnatal rupture common
4 Bacterial infection may complicate either ruptured or unruptured aneurysms of the aortic sinus
5 *Cardiac catheterization in cases wherein aneurysm communicates with right side of heart shows* high oxygen content in blood of right ventricle alone or in right ventricle and right atrium depending on location of communication

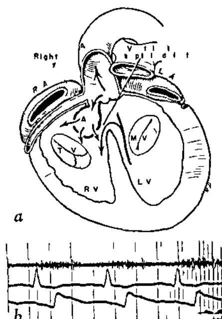


Fig 195—a The intracardiac circulation in the malformation of the patient whose heart is illustrated in Figures 194 196 and 198 *b* The phonocardiogram E C G and carotid pulse



Fig 196—The heart and lungs of the patient whose heart is illustrated in Figures 194 and 198 There is marked cardiac enlargement The pulmonary trunk (P T) is markedly dilated explaining the roentgenographic contour of the heart as shown in Figure 197

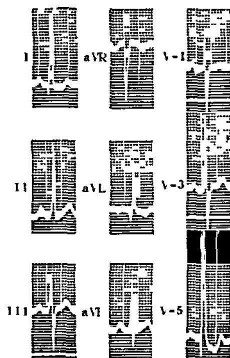




Fig 197—Electrocardiogram and the roentgenogram of the thorax of the patient whose heart is illustrated in Figures 194 196 and 198

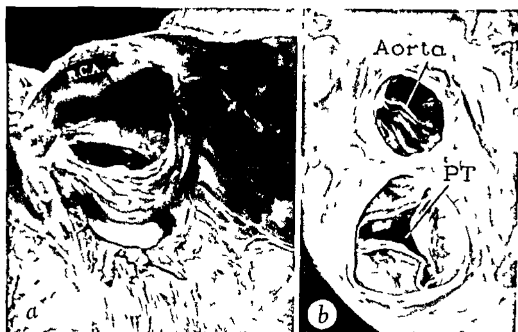


Fig 198—From the patient whose heart is illustrated in Figures 194 and 196 *a* A portion of the aortic valve is illustrated Beneath the ostium of the right coronary artery (R C A) is shown a portion of the mouth of the aneurysm of the right aortic sinus Beneath the right aortic cusp is the crescent shaped ventricular septal defect which is illustrated from the right side in Figure 194 *b* By virtue of the greatly excessive pulmonary blood flow the pulmonary trunk is dilated The aorta is of normal caliber P T = the pulmonary trunk immediately above valve level Aorta = the ascending aorta immediately above valve level

Synopsis of Cardiac Catheterization Data

Origin of blood sample site of pressure recordings	Pressure mm Hg		Van Slyke analysis			Per cent oxygen saturation (whole blood oximeter)
	Systolic	Diastolic	O content	O capacity	Per cent saturation	
Right atrium	14	0	14 2	20 5	69 3	67 5
Right ventricle inflow tract	78	10	16 8	20 9	80 4	78 0
Right ventricle outflow tract	67	9	19 0	0 6	9[illegible]	94 0
Pulmonary trunk	54	21				9[illegible] 5
Left pulmonary artery	60	23	18 2	20 4	89 [illegible]	90 [illegible]
Right radial artery	133	54	19 5	20 4	95 6	94 5

Subaortic Stenosis

SUBAORTIC STENOSIS is characterized by the presence of a ring of muscular and fibrous tissue immediately below the aortic valve. It probably represents the incomplete evolution of the bulbus cordis. It has the same effect on the heart as an acquired aortic stenosis and produces signs and symptoms which may be impossible to differentiate from acquired aortic stenosis although the aortic second sound is expected to be preserved in subaortic stenosis. Cardiac failure and subacute bacterial endocarditis are the common complications. Survival to adult life is frequent.

Subaortic Stenosis

IN THIS heart there is a localized zone of stenosis in the outflow tract of the left ventricle The edges of the stenotic zone are rimmed by fibrous tissue which adds greatly to the degree of narrowing As a consequence of the obstruction there is hypertrophy of the left ventricular wall The leaflets of the aortic valve in this specimen are deformed as a result of healed bacterial endocarditis (penicillin treated)

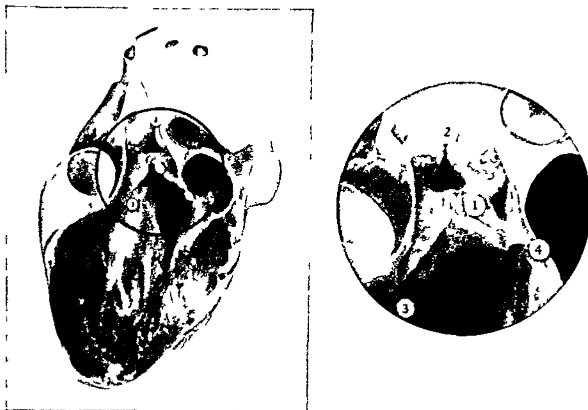


Fig 199—Model of heart *a* x½ Interior of left ventricle and ascending aorta (1) Fibrous tissue causing subaortic stenosis (2) Aortic valve (3) Ventricular septum (4) Anterior leaflet of mitral valve *b* Close up view of subaortic region and aortic valve of model illustrated in *a*

History of the Patient

MAN 37 years old There was a questionable history of rheumatic fever when 6 years old leakage of the heart known since the age of 12 years increasing dyspnea on exertion since the age of 35 years One year later was hospitalized because of dyspnea and cough but no dependent edema The condition deteriorated requiring digitalis and salt restriction Since first hospitalization he had recurrent episodes of fever for which antibiotics were given When seen three months prior to death there was clinical evidence of subacute bacterial endocarditis Examination revealed aortic systolic and diastolic murmurs marked cardiac enlargement with mild congestive heart failure The electrocardiogram revealed evidence of left ventricular hypertrophy Treated with antibiotics with temporary improvement On rehospitalization with congestive heart failure the patient died suddenly

Principal Clinical Features of This Anomaly

1 Systolic murmur over the aortic area propagated along cervical vessels present since early childhood
2 Varying degrees of left ventricular hypertrophy by roentgenogram
3 Evidence of left ventricular hypertrophy by electrocardiogram
4 Congestive cardiac failure and bacterial endocarditis are frequent complications





Fig 200—Specimen from which models shown in Figure 199 were prepared *a* The outflow tract of the left ventricle Accentuating the narrowing caused by prominence of the ventricular septum is a collar of fibrous tissue attached in part to the anterior leaflet of the mitral valve (M V) and to the muscular part of the ventricular septum (V S) *b* The outflow tract of the left ventricle and the aortic valve opened Lying above the fibrous collar (F C) is the aortic valve The posterior leaflet of the aortic valve (P) is thickened by a process of healed bacterial endocarditis M V = anterior leaflet of the mitral valve V S = muscular portion of the ventricular septum

I V-1 II V-3 III V-5

Fig 201—The electrocardiogram and the thoracic roentgenogram of the patient whose [illegible] strated in F[illegible] and 200

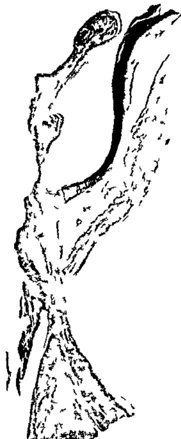




Fig 03—(above)—The opened aortic valve and subaortic collar in a case of sub aortic stenosis in a ?5 year old man with bacterial endocarditis involving the sub aortic collar and the aortic valve There is an aneurysm of the ventricular septum inferior to the right aortic cusp (From Morrison R W and Edwards J E *J Tech Methods 31* 73 1950 with permission)

Fig 02—(left)—Photomicrograph of the posterior aortic leaflet and the subaortic fibrous collar in the case of subaortic stenosis illustrated in Figures 199 200 and ?01 The irregular thickening of the cusp is the result of healed bacterial endo carditis (x5)



Fig 204—(left)—Subaortic stenosis and a patent ductus arteriosus from another 25 year old man who died of congestive card ac failure The aortic valve is viewed from above The cusps have been retracted laterally exposing the subaortic fibrous collar (From Morrison R W and Edwards J E *J Tech Methods 31* 73 1950 with permis sion)

Atresia of the Aortic Orifice

(*Functional Two chambered Heart*)

Atresia of the aortic orifice is characterized by fusion of the aortic leaflets to form an imperforate diaphragm at the level of the aortic valve The ventricular septum is intact In some cases aortic atresia and mitral atresia may coexist Most cases however have a small but normally developed mitral valve Whether or not mitral atresia co exists the normal outlet for the left side of the heart is closed and the blood is shunted from the left atrium to the right usually through a patent foramen ovale The right atrium and right ventricle thus be come in essence a common atrium and a common ventricle respec tively The systemic circulation is supplied through the pulmonary artery by way of a patent ductus arteriosus

Survival beyond early infancy is rare There is a particular tendency for this malformation to occur in males

Atresia of the Aortic Orifice

(Functional Two chambered Heart)

In this heart the primary malformation is atresia of the aortic orifice

The ductus arteriosus and the foramen ovale are patent and the right atrium right ventricle and pulmonary trunk are enlarged The left ventricular wall is thick but its chamber is diminutive

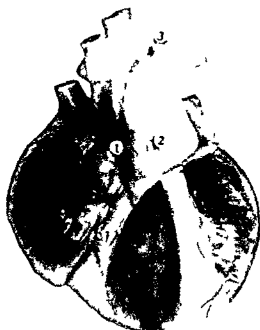


Fig 205—Anterior view (model x1) (1) Site of atretic aortic orifice (2) Dilated pulmonary trunk (3) Patent ductus arteriosus



Fig 206—Left anterior view (model x1) (1) Diminutive left ventricular chamber (2) Thick left ventricular wall

History of the Patient

Girl 3½ months old intermittent cyanosis occurring with crying and exertion began at the age of 2 weeks The infant did not gain weight Dyspnea and cyanosis became more intense A short basal systolic murmur was present Hemoglobin amounted to 19 gm per 100 cc of blood erythrocytes numbered *5 670 000 per cubic millimeter of blood Thoracic roentgenograms showed right ventricular enlargement* with prominent pulmonary arterial shadow Electrocardiogram showed right axis deviation The infant died suddenly

Principal Clinical Features of This Anomaly

1 Progressively severe cyanosis from birth
2 Infant is usually very dyspneic and weak
3 Early development of cardiac failure
4 Systolic murmur usually present
5 Roentgenologic aspects marked enlargement of right ventricle and pulmonary artery
6 Electrocardiogram right ventricular hypertrophy
7 Death in early infancy

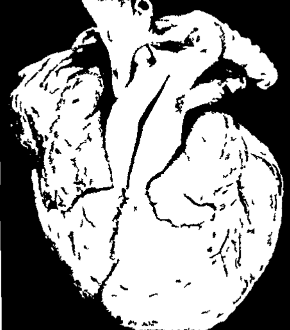


Fig 207—Exterior of heart and great vessels showing greatly dilated pulmonary trunk and hypoplastic ascending aorta

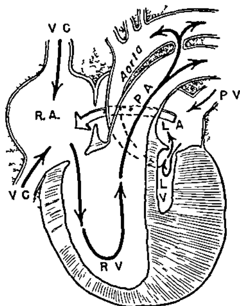


Fig 208—Diagram of intracardiac circulation in atresia of aortic orifice

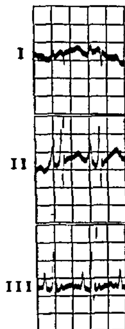




Fig 209—Electrocardiogram and thoracic roentgenogram of the patient whose heart is shown in Figures 205 207



Fig 210—Specimen from which models in Figures 205 and 206 were prepared *a* The great vessels have been opened showing the large pulmonary trunk the patent ductus *arteriosus and the hypoplastic ascending aorta* *b* The left side of the heart Though the chamber of the left ventricle is diminutive the wall is greatly hypertrophied Compare this feature with the right side of the heart in pulmonary atresia with intact septum (Fig 148 page 94) There is endocardial thickening of the left atrium A probe lies in the patent foramen ovale

Atresia of the Aortic Orifice in a 5-day-old boy

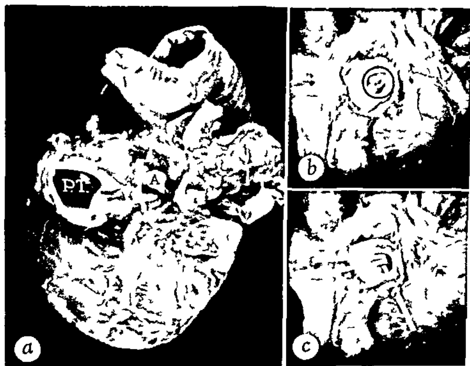


Fig 211—*a* Superior aspect of the heart showing dilated pulmonary trunk (P T) and hypoplastic ascending aorta (A) *b* The aortic valve (within circle) from above The valve is atretic on the basis of diaphragm like fusion of the valvular tissue *c* The same view as shown in *b* with the addition of probes *in the ostia of the coronary arteries showing that the coronary arteries* arise superior to the atretic valve These vessels are supplied in retrograde fashion from the ascending aorta (see Fig 208) (Fig 211*b* from Baggenstoss A H *J Tech Methods* 20 62 1940 with permission)

Coarctation of the Aorta

COARCTATION of the aorta is a malformation characterized by a deformity of the aortic media causing narrowing usually severe of the aortic lumen The pathological and clinical features depend to a great extent on the relationship of the coarctation to the aortic entrance of the ductus arteriosus and also on whether or not the ductus arteriosus is closed In the majority of instances the coarctation lies beyond the ductus and the ductus obliterates normally In this type of case there is no abnormality of the pulmonary circulation A well developed collateral circulation by passes the coarctation

In the cases associated with a patent ductus arteriosus there may be pulmonary hypertension In the usual case of coarctation of the aorta associated with a closed ductus arteriosus the patient lives to adult life although life expectancy is materially reduced Among the causes of death are failure of the left ventricle rupture of the aorta bacterial endocarditis of the commonly associated bicuspid aortic valve and rupture of an aneurysm of the circle of Willis

Coarctation of the Aorta

In this aorta there is coarctation just beyond the ligamentum arteriosum In the external view there is a characteristic concavity involving the anterior superior and posterior aspects of the aorta The lower aspect of the aorta that receiving the ligamentum does not share in the concavity The narrowing of the lumen however is much greater than might be judged from the external appearance (see Fig 213 also)

Fig 212—Model of heart and aorta showing coarctation *(left)* External view (1) Depression of superior aspect of aorta at location of coarctation ( ) Ligamentum arteriosum *(right)* Close up view of interior of aorta in the region of coarctation showing infolding of aortic wall causing marked narrowing of lumen Lesion modeled after specimen region Arrow in aortic lumen at coarctation A = descending aorta



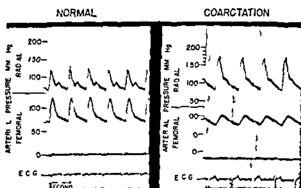


Fig 213—(left)—Longitudinal section of aorta removed surgically by Dr O T Clagett Beyond the ligamentum arteriosum (Lig art) the superior aspect of the aortic wall shows characteristic infolding of the media causing narrowing of the aortic lumen Accentuation of narrowing results from superimposed fibrous intimal thickening in this region Arrow in aortic lumen at coarctation A = descending aorta

Fig 214—(right)—Pulse waves and pressure recordings of radial and femoral arteries in a normal patient and in a patient with typical findings of coarctation of the aorta

The patient a 19 year old woman was first seen in December 1947 Hypertension known since 12 years of age no symptoms except mild intermittent headaches Blood pressure in left arm 180/120 and in right arm 176/116 Systolic murmur at base of heart transmitted to neck Diminished pulses in abdominal aorta and femoral arteries Resection of coarctation and end to end anastomosis of aorta January 7 1948 Post operatively significant fall in blood pressure in arms One and a half years later the patient was delivered of a healthy baby without incident

Fig 215—Preoperative electrocardiogram and roentgenogram of thorax Rib removed during operation showing characteristic location of notching Other illustrations on this case appear in Figures 212 213 and 216

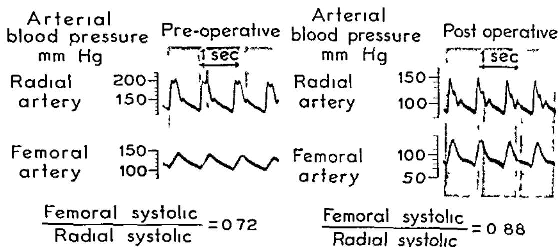


Fig 216 Preoperative (*left*) and postoperative (*right*) arterial blood pressure recordings in radial and femoral arteries

Coarctation of the Aorta with Patent Ductus Arteriosus
Coarctation of the Aorta Proximal to the Left Subclavian Artery

TWO INSTANCES of coarctation of the aorta are shown here In one the coarctation is proximal to a patent ductus arteriosus In the other the constriction is in an atypical position between the origins of the left sub clavian and the left common carotid arteries while the ductus arteriosus is closed



Fig 217—Coarctation proximal to patent ductus arteriosus (model x1) (1) Site of aortic coarctation (2) Patent ductus arteriosus



Fig 218—Unusual type in adult (half size model) (1) Site of aortic coarctation (2) Left subclavian artery (3) Ligamentum arteriosum

History of These Patients

A BOY 3 days old (Fig 217) apparently was normal at birth but on the third postnatal day became cyanotic and died suddenly

A MAN 26 years old (Fig 218) was *admitted with cardiac failure Aortic systolic and diastolic murmurs* and cardiac enlargement Blood pressure in the right arm was 210 systolic and 40 diastolic in the left arm it was 110 systolic and 78 diastolic It was not obtainable in the legs Pulse could not be felt in the abdominal aorta and femoral arteries A thoracic roentgenogram showed cardiac enlargement and absence of the aortic knob but no notching of ribs Patient died of a bacterial endocarditis involving a bicuspid aortic valve

The case illustrated in Figure 218 reported by Parker R L and Dry T J *Am Heart J* *15* 739 1938

Coarctation of the Aorta with Unequal Blood Pressure in Arms

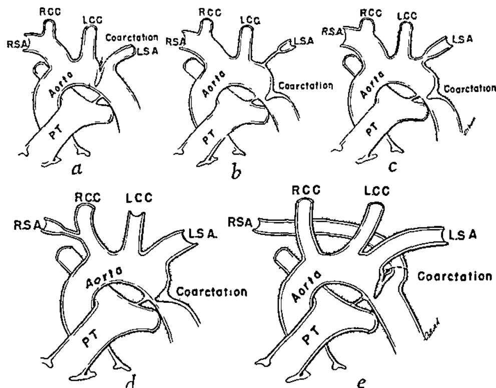


Fig 219—Variations in structure leading to unequal blood pressure in the arms in coarctation of the aorta *a* Coarctation proximal to left subclavian artery as in case illustrated in Figure 218 *b* Coarctation in usual location but with atresia of origin of left subclavian artery (as in the case of Woltman H W and Sheldon W D *Arch Neurol & Psychiat* 17 303 1927) *c* Coarctation in usual location but with stenosis of left subclavian artery (as in the case of Bauer D deF and Iverson L *Am Heart J* 30 30 1945) *d* Coarctation in usual location associated with stenosis or atresia of right subclavian artery (as in the case of Lowe W S and Holmes J H *Am Heart J* 17 678 1939) *e* Coarctation in usual location associated with right subclavian artery arising independently from the aorta as its fourth branch (as in case 12 of Fawcett J *Guy's Hosp Rep* 59 1 1905) In conditions illustrated in *d* and *e* the systolic blood pressure is low in the right arm and in *a* *b* and *c* it is low in the left arm

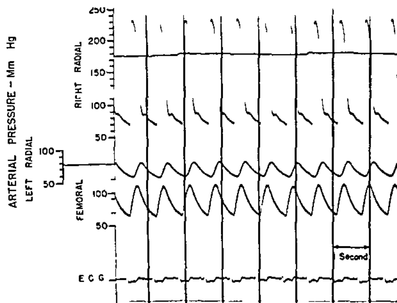
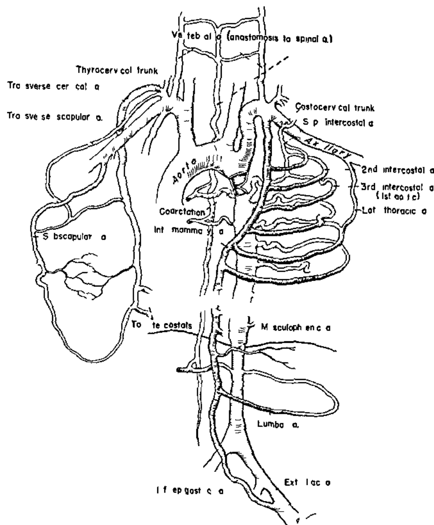


Fig 220—Arterial pressures and pulse tracings in a case of coarctation of the aorta with unequal blood pressures in the arms The left radial pressure is of a level comparable to the femoral while the right radial systolic pressure is markedly elevated Pulse contours of the left radial and femoral arteries are similar to each other and each is dissimilar to contour in right radial artery It is postulated that this patient has a malformation of a type illustrated in Figure 219*a* *b* or *c* (From Burchell H B *et al* *M Clin North America* 34 1177 1950 with permission)

Collateral Circulation in Coarctation of the Aorta



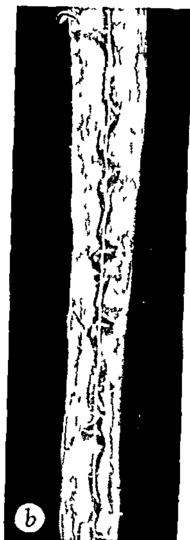
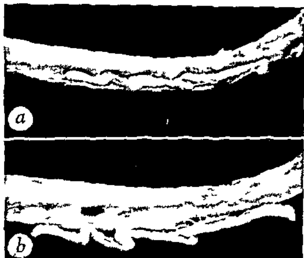


Fig 21 a—Diagrammatic representation of routes of collateral circulation in coar tation of the aorta *b* Dilated tortuous anterior spinal artery representing a collateral channel in a 28 year old man with coarctation of the aorta



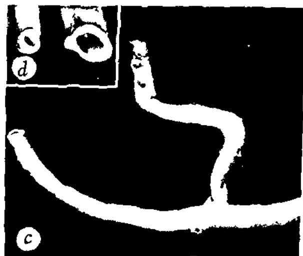


Fig 22 —a A rib and intercostal artery from an 18 year old man with coarctation of the aorta *b* The same specimen as illustrated in *a* but with the artery retracted to show the eros on of the rib and serration of the costal groove created by the tortuous points in the ves el *c* The external iliac artery and inferior epigastric artery in a 13 year-old boy with coarcta tion of the aorta Distal to the entrance of the inferior epigastric artery (tortuous vessel) which itself is dilated and acting as a collateral channel the external iliac artery dilat s indicating that substantial blood is carried to the leg by way of the inferior epigastric *d* The up turned proximal (*left*) and distal (*right*) ends of the ext rnal iliac artery illustrated in *c*

Coarctation of the Aorta, Complicating Lesions



Fig 23a—Bicuspid aortic valve in a 28 year old man with coarctation of the aorta Arrow points to focal fibrous lesion on anterior leaflet of mitral valve interpreted as the reaction to a regurgitant stream of blood Other illustrations on this case appear in Figures 1*b* and 225*b*



Fig 23b—Bicuspid aortic valve complicated by bacterial endocarditis in a man 25 years old with coarctation of the aorta



Fig 24—Aortic coarctation in a man 29 years old The probe runs through the zone of coarctation Tip of probe points to corrugated patch in distal portion of aorta interpreted as a jet lesion



b

Fig ? 4b—Photomicrograph of a portion of the corrugated patch and adjacent aorta shown in *a* Lesion consists of loss of medial tissue and presence of intimal fibrosis (Verhoeff's elastic tissue stain counterstained with van Gieson's connective tissue stain x1)

Aortic arch
An
Descending aorta
Lig art
a
b
c

Fig 225—a Segment of aorta and aneurysm of intercostal artery removed surgically by Dr O T Clagett from a woman 24 years old The coarctation lies just distal to the aortic entrance of the ligamentum arteriosum (Lig art) The lumen of the aneurysm (An) of the intercostal artery is continuous with the lumen of the descending aorta *b* Circle of Willis with two so called congenital aneurysms in the 28 year old man with coarctation on whom other illustrations appear in Figures 221*b* and 223*a* *c* Left ventricle and aorta in a 34 year old woman with coarctation of aorta (within smaller circle) who died of hemopericardium complicating dissecting aneurysm of the aorta Tear in ascending aorta within larger circle (Illustration submitted by Dr Timothy Leary)

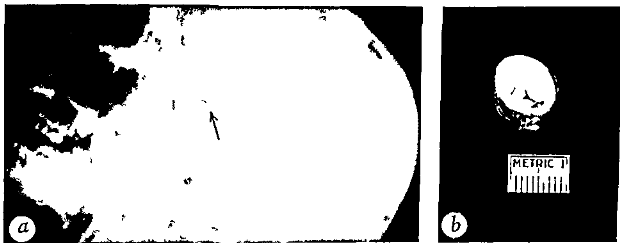


Fig 226a—Cerebral angiogram in a 20 year old man with coarctation of the aorta showing an aneurysm of the circle of Willis (tip of arrow) On December 6 1950 ligation of the right internal carotid artery for recurrent subarachnoid hemorrhage The aortic segment containing the coarctation was resected and end to end anastomosis of the aorta was per formed on February 15 1951 *b* The segment of aorta removed surgically The distal end of the specimen is illustrated showing a high degree of luminal narrowing Postoperatively there was reduction of brachial blood pressure and estab lishment of forceful femoral arterial pulsations Patient s condition currently good

Coarctation of the Aorta in an Infant, Surgical Correction

GIRL 8 weeks old Feeding difficulty several weeks after birth No cyanosis or murmurs Cardiac enlargement Absent femoral arterial pulsations Electrocardiogram right ventricular hypertrophy Aortogram coarctation proximal to left subclavian artery Operation by Dr J W Kirklin at 10 weeks of age resection of coarctation and portion of left subclavian artery end to end anastomosis of aorta Postoperatively femoral pulsations palpable Subsequent growth and development of patient good

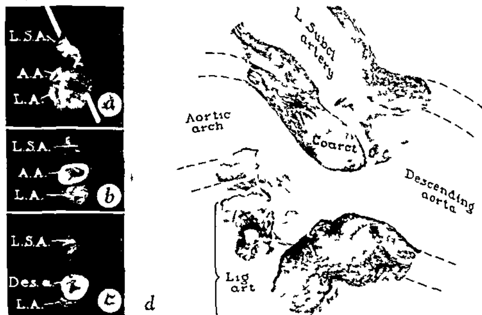


Fig 27a b c and d—The segment of aorta and the left subclav an artery removed surgically *a* The specimen from in front *b* The specimen from below *c* The specimen from above L S A = left subclavian artery A A = aortic arch L A = ligamentum arteriosum Des a = d scending aorta In *a* the probe extends through the left subclavian artery into the descend ng aorta *d* Photomicrograph of the surgical specimen of aorta prepared in the plane ill st ted in *a* The coarctation (Coarct) lies prox mal to the left subclavian artery There is stenosis of the mouth of the left subcl i n artery

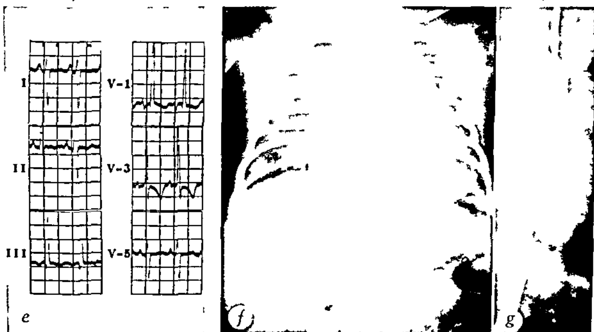


Fig 2 7e f nd g—Preoperative stud es electrocard ogram roentgenogram of the thorax and aortog am Aortography performed by Dr D G Pugh Rad op que dye was injected retrograde into left brachial artery

Coarctation of the Aorta with Left Ventricular Failure at 7 Weeks of Age

GIRL 7 weeks old Apparently normal at birth feeding difficulty from birth Became acutely ill with high fever respiratory distress and increasing cyanosis Heart enlarged but no murmurs Despite administration of oxygen and of antibiotics patient died the day after admission (From Bahn R C *et al* *Pediatrics* 8 192 1951 with permission)

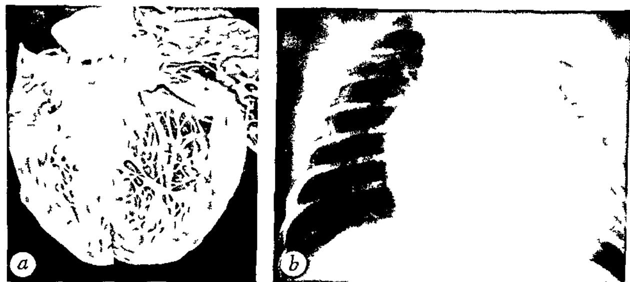


Fig 228a and b—a Dilated and hypertrophied left ventricle Bicuspid aortic valve *b* Roentgenogram of the thorax

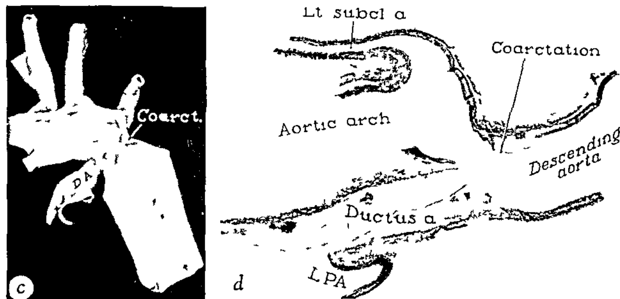


Fig 228c and d—c Gross specimen of aorta Opposite aortic entrance of the closing ductus arteriosus (*D A*) is an infolding of the aortic wall (Coarct) characteristic of aortic coarctation The intercostal arterial ostia are of approximately normal width suggesting an inadequate collateral system *d* Photomicrograph of a portion of the specimen illustrated in *c* showing the coarctation opposite the aortic mouth of a closing ductus arteriosus L P A = left pulmonary artery

Coarctation of the Aorta Distal to Patent Ductus Arteriosus

FEMALE 15 years old observed in 1945 Harsh systolic murmur at apex and pulmonary area Intermittent cyanosis of left hand Electrocardiogram right axis deviation Death following operation for scoliosis (From Edwards J E *et al* *Am Heart J* 38 205 1949 with permission)

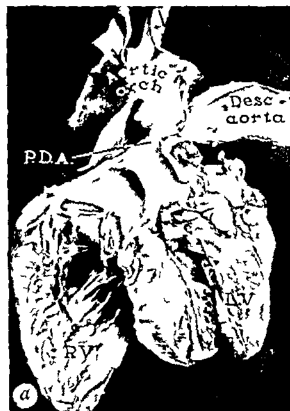


Fig 229a—The heart pulmonary al e and great es el Patent ductus arteriosus (P D A) oppos te left subcla ian artery and proximal to aortic coarctat cn (ontaining probe) Hypertrc phy of ventricular walls



Fig 29b—Pulmonary muscular artery Mod rate medial hypertrophy elast c fragmentation and pronoun ed ntimal fibrous thicken ng Appreciable narrowing of lumen

Coarctation Proximal to Patent Ductus Arteriosus

GIRL 7 years old Harsh apical systolic murmur continuous murmur over pulmonary area thrill over suprasternal notch Blood pressure in arms 138 150/45 50 Blood pressure in left leg 90 102/50 60 Femoral pulses forcible No clinical or pathologic evidence of collateral circulation No cyanosis in any part (From Edwards J E *et al* *Am Heart J* 38 205 1949 with permission)





Fig 30—a H rt ante ior v ew Hypert ophy of vent icles *b* A no mal pulmonary musc la rtery from a 6-yea old ch ld show ng th w ll a d wide lumen P lmonary mu cul r artery f om the p t ent who e he rt s llu trated in *a* Th ck muscular med a No nt mal thick n ng P onounced l m l nar owing

Coarctation of the Aorta with Turner's Syndrome

ONE OF the rare syndromes in which there exist associated anomalies has been named after Turner The main features are an anomalous somatic development with ovarian agenesis The most prominent of the body malformations is the short neck with marked weblike folds of the skin on either side There is also an increased carrying angle of the elbows and widely spaced incisor teeth Dwarfism is present and is usually causally related to the lack of ovarian function Coarctation of the aorta is not an infrequent associated anomaly and for that reason the syndrome seems to earn a place in this section of the atlas on coarctation of the aorta



Fig 231—Patient with coarctation of the aorta and Turner s syndrome

History of the Patient

A GIRL 9 years of age was brought to the clinic for evaluation of a cardiac murmur The murmur was basal in type and of moderate intensity and the outstanding finding on cardiovascular examination was the near absence of femoral pulsations The diagnosis of coarctation of the aorta was made It was noted that there was marked webbing of the skin of the lateral aspects of the neck the upper incisor teeth were spaced and the elbows had an increased carrying angle The general development and body build for her age group were otherwise normal Analyses for gonadotropin excretion were not carried out Plastic surgery for the neck was recommended and performed but it was recommended that repair of the coarctation be deferred until she would be a few years older

Arachnodactyly

(Marfan's Syndrome)

AMONG the various syndromes wherein congenital anomalies of the heart may be associated with other multiple somatic deformities (or congenital organ weakness) Marfan's syndrome is one of the most interesting In its complete form there is the tall thin gracile habitus with the long spindly fingers and toes poor muscular development and bilateral dislocation of the lenses The cardiac defect is sometimes a patent foramen ovale No obvious cardiac defect may be present yet the patient has an occult vascular weakness which may result in aortic rupture or dilatation of the aorta associated with aortic valvular in sufficiency Incomplete forms of the syndrome may be recognized The ophthalmologists in particular have been especially active in drawing attention to these cases especially when the complete Marfan syndrome with ectopia lentis is present

Some of the less constant manifestations of the condition are dolicho cephaly with a narrow palatal arch contractions and subluxations of joints a high position of the patella kyphosis and scoliosis and either a funnel depression or pigeon breast deformity of the thorax

Arachnodactyly

(Marfan's Syndrome)

In 1943 a woman 34 years old while putting on her coat suddenly complained of substernal pain and inability to breathe Dead on the arrival of the physician At the Mayo Clinic the diagnosis of arachnodactyly made in 1932 No cardiac lesion was clinically diagnosed Death from hemopericardium complicating dissecting aneurysm of aorta (Case reported by Dvorak H J *Proc Staff Meet Mayo Clin* 7 715 1932 and by Burch F E *Arch Ophth* 15 645 1936 Case 4)

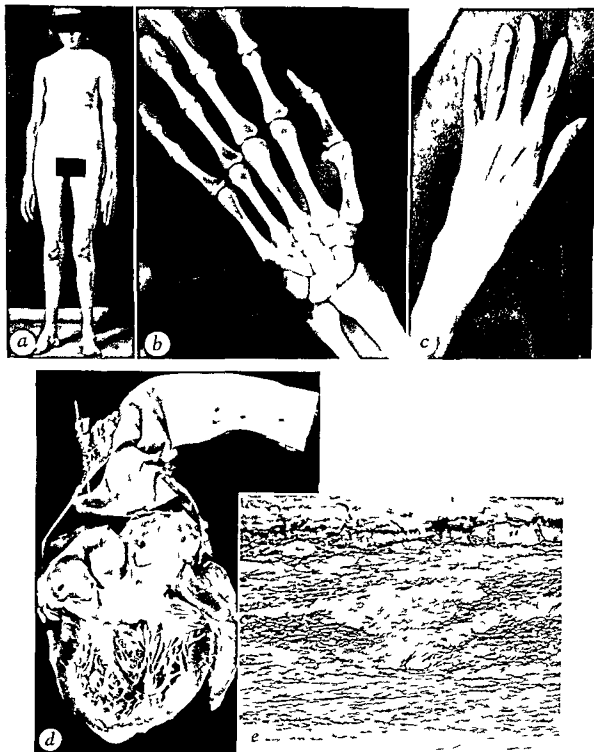


Fig 232—*a* The patient *b* Roentgenogram of left hand *c* Photograph of left hand *d* Left ventricle and aorta Horizontal tear of ascending aorta with dissecting aneurysm Probe extends from intramural hematoma of aorta into that extending to innominate artery *e* Photomicrograph of aorta showing focal deficiency in elastica of media

Vascular Rings

VASCULAR RINGS are anomalies of the aortic arch system A vascular ring is of consequence if it interferes with the function of the trachea or the esophagus it may result in death if unrecognized and if the obstruction is not relieved surgically

The commonest types of vascular rings are (1) anomalous origin of the right subclavian artery as the fourth branch of an otherwise normal aorta to reach its destination it must cross the midline behind the esophagus from left to right (2) functioning double aortic arch and (3) single functioning right or left aortic arch passing behind esophagus to reach descending aorta which is on the contralateral side

Anomalous Aorta Double Aortic Arch

DOUBLE AORTIC ARCH with a left sided ligamentum arteriosum is illustrated in this diagram The right arch after passing over the right bronchus crosses to the left behind the esophagus to join the upper end of the descending aorta which is on the left side

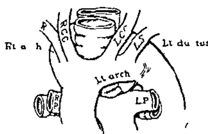


Fig 233—Double aortic arch in which the two arches are of equal size Ligamentum arteriosum and descending aorta are on the left (From Edwards J E M *Clin North America* 32 925 1948 with permission)

History of the Patient

FEMALE 6 weeks old noisy breathing since the age of 1 week At the age of 4 weeks an enlarged thymus was thought to be present for which radiation therapy was given Following this respirations became noisier coughing became persistent and slight cyanosis developed No cardiac murmurs

Roentgenoscopic examination revealed a constriction of the esophagus Exploration of the thorax revealed a double aortic arch in which the two arches were of equal diameter The right arch and the ligamentum arteriosum were divided

The patient died on the third postoperative day

Principal Clinical Features of Vascular Rings

1 Dysphagia (dysphagia lusoria) may or may not be present
2 In infants there may be marked respiratory distress with stridor and inspiratory retraction of intercostal and supraclavicular tissues
3 In adults extrinsic pulsation has been seen posteriorly on esophagoscopy
4 Roentgenoscopic examination reveals evidence of right aortic arch anomalous subclavian artery or double aortic arch
5 Usually there is no evidence of intracardiac anomaly



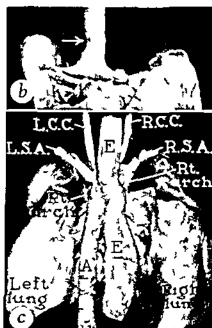


Fig 34a b and c—From the case described on preceding page *a* Roentgenogram of the thorax *b* The trachea and major bronchi There is indentation of the trachea (point of arrow) caused by pressure of the right aortic arch against the trachea *c* Thoracic organs from behind The right arch had been divided surgically near its junction with the descending aorta The ends of the divided right arch have retracted exposing the esophagus (E) where it had been compressed R S A = right subclavian artery R C C = right common carotid artery L C C = left common carotid artery L S A = left subclavian artery A = descending aorta





Fig 34d and e—Esophagrams revealing pronounced narrowing of the esophagus in the upper part of the thorax

Anomalous Aorta Left Aortic Arch with Right Descending Aorta and Right Ligamentum Arteriosum

This anomaly which is rare is the mirror image of a commoner type of anomaly In the latter the aortic arch passes over the right bronchus and then behind the esophagus to join the left sided descending aorta In this specimen however the aorta passes over the left bronchus and then courses to the right behind the esophagus to join the descending aorta which is on the right side The ligamentum arteriosum is on the right extending from the right pulmonary artery to the aorta The right subclavian artery arises from the aorta at the junction of the aortic arch and descending aorta There is no intracardiac anomaly



Fig 235—Anterior view (model x1) (1) Aortic arch (2) Trachea and esophagus (3) Pulmonary trunk



Fig 236—Viewed from above (model x1) (1) Trachea and esophagus encircled and compressed by vascular ring (2) Right sided descending aorta (3) Right sided ligamentum arteriosum

History of the Patient

Male 17 months old had had an imperforate anus since birth, dysphagia noted since 1 year of age colostomy performed shortly after birth for latter condition but progress was poor The infant died of intestinal obstruction This malformation is related to the double aortic arch illustrated in Figure 241c (Case reported by Edwards J E *Proc Staff Meet Mayo Clin 23 108 1948*)

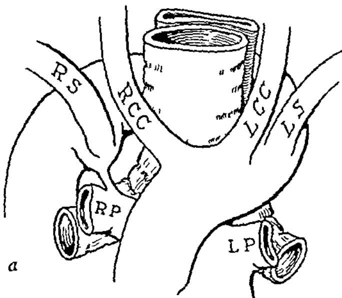


Fig 237a—Drawing of the specimen illustrated in Figures 235 36 237*b* and 238 There is a left aortic arch which crosses behind the esophagus to join a right sided descending aorta There is a diverticulum representing a posterior remnant of the right arch at the junction of the left arch and of the descending aorta From this diverticulum arises the right subclavian artery and the ligamentum arteriosum The latter is right sided and is inserted into the diverticulum

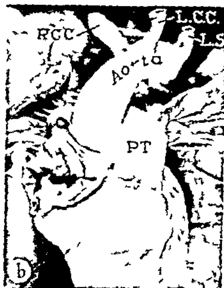


Fig 237b—The heart lungs and great vessels from the front The ascending aorta passes into a left sided aortic arch RCC = right common carotid artery LCC = left common carotid artery LS = left subclavian artery PT = pulmonary trunk

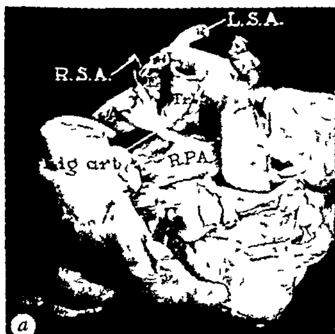


Fig 238—The specimen illustrated in Figures 235 237 viewed from a right superior angle The esophagus (E) and trachea (Tr) are encircled by a vascular ring composed of the left aortic arch on the left the retroesophageal portion of the aortic arch behind the right sided ligamentum arteriosum (Lig art) on the right and the right pulmonary artery (R P A) in front

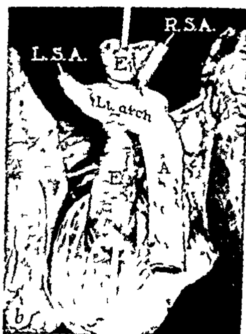


Fig 238b—The mediastinal structures from behind The left aortic arch passes from left to right behind the esophagus to join the right sided descending aorta (A) E = esophagus LSA = left subclavian artery RSA = right subclavian artery

Anomalous Aorta Origin of Right Subclavian Artery as a Fourth Branch of Otherwise Normal Aorta

A 63 year old man with carcinoma of the bladder No symptoms of esophageal obstruction





Fig 39a and b—Specimen from the patient whose esophagrams are illustrated in *c d* and *e* *a* Posterior view of great vessels trachea and esophagus The right subclavian artery arises as the fourth branch of the aorta and passes to the right behind the esophagus *b* The trachea esophagus and aorta viewed from above The esophagus is compressed as it lies between the aorta on the left and the right subclavian artery behind R C C = right common carotid artery L C C = left common carotid artery L S A = left subclavian artery Tr = trachea E = esophagus







Fig 239c d and e—Esophagrams from case illustrated in *a* and *b* Distortion of esophageal contour by anomaly

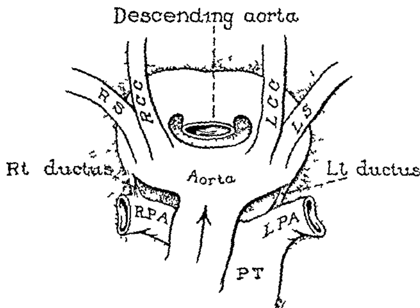


Fig 40—This diagram represents a hypothetical anomaly It consists of a double aortic arch with double ductus arteriosus The descending aorta is in a neutral position This form may be considered the parent form from which all the known and hypothetical anatomic varieties of malformations of the a rti syst m may be diagrammatically derived By loss of one ductus arteriosus and by the shift of the descending ao ta to the r ght or to the left four bas c patterns of double aorti arch may be d rived These are illustrated in Figure 241 (From Edwards J E pp r 503 in Gould S E *Pathology of the Hea t* Charles C Thomas Publisher Springfield Illinois, 1953 with p rm ssion)

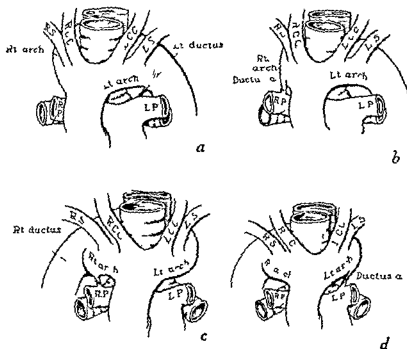


Fig 241—F r variations of double aorti arch Ea h may be onsidered a basic patt rn from which other *forms of mal formations of the aorti arch may be derived* In each case other anatomic variet es of *malformations* devel p either by narrow ng r by loss of erta n elements of the double aortic *arch* *a* Double aortic arch with left sided descending aorta nd left sided ductu a teri sus *b* Double aortic arch w th left sided descend ng aorta and r gl t s ded ductu arte osus D bl ao ti arch w h r ght sided descending aorta and right sided ductus arter o us *d* Double ort c arch w th right d descending aorta and left sided d ctu rteriosus (F g 241*a* and *c* fr m Edwards J E *M Cl n North A er a* 3 9 5 1949 with perm ssion Fig 1*1b* and *d* from Kirkl n J W and Clagett O T *Pr c Staff Meet Mayo Clin* 5 360 1950 with perm ssion)

Origin of Left Coronary Artery from Pulmonary Trunk

THE LEFT coronary artery in this heart arises from the pulmonary trunk while the right coronary artery arises in a normal manner from the aorta The left ventricle is dilated and there is *scarring of the myocardium* in the distribution of the anomalous left coronary artery



Fig 243a—Right ventricle and pulmonary trunk showing origin of left coronary artery (point of arrow) from pulmonary trunk



Fig 243b—Left ventricle and aorta showing origin of only right coronary artery (point of arrow) from aorta (Specimen submitted by Dr Frederic Parker Jr)

History of the Patient (History supplied by Dr James M Baty)

FEMALE first seen at 3½ months of age because she did not eat well and at times had rapid and grunting respirations during feeding No murmurs were heard liver and spleen enlarged Respiratory distress increased Feedings interrupted by dyspnea Heart rate rapid and rhythm of gallop quality Cardiac size *increased progressively* a *barium swallow did not reveal any* displacement of esophagus Blood counts normal Digitalization resulted in temporary improvement but gallop rhythm persisted Apical systolic murmur heard for first time at age of 5 months Dyspnea tachycardia and irritability recurred at age of 6 months and regurgitation became more frequent At 8 months of age extreme respiratory distress and cyanosis developed and patient died enroute to hospital

Principal Clinical Features of This Anomaly

1 Feeding (which represents exertion) becomes *difficult infant may cry draw up legs and display* discomfort sweating dyspnea or grunting respirations Feeding may be followed by shocklike state Malnutrition common
2 *Progressive cardiac enlargement* associated with gallop rhythm and eventually congestive heart failure which responds poorly to treatment Sudden death occurs
3 Murmurs are absent except for systolic apical murmur probably related to cardiac dilatation
4 Cyanosis is absent except in terminal stages
5 Roentgenologic examination reveals progressive *cardiac enlargement (mostly left ventricular dilatation)*
6 Electrocardiographic changes at times resemble those seen in adults with myocardial ischemia Tachycardia is usual and the ventricular complexes may have a low amplitude Evidence of right ventricular hypertrophy common in other forms of congenital malformation of the heart is seldom seen
7 Death in infancy is common but a few patients reach adult life



Fig 44—Left s de of heart illustrated in Figure 243 Dilatation of left atrium and left ventricle Scarring of left ventri ular wall Secondary endocardial sclerosis

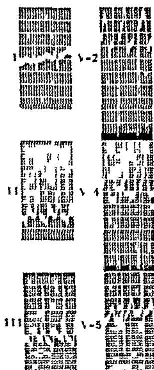




Fig 4 —El tr rd gram and r en g nogram of the thor f the p ti nt wh se heart i illu trated in Figures 43 n l 44 (Illustr t n suppl ed by and r produ ed with per n s on cf Dr Edward F Bland)

Origin of Right Coronary Artery from Pulmonary Trunk

In this heart the right coronary artery arises from the pulmonary trunk. The left coronary artery arises from the aorta

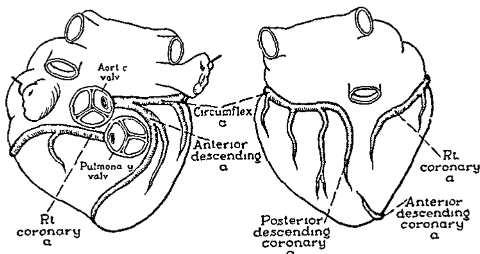


Fig 46—Distribution of the left and right coronary arteries The right coronary artery arises from the right sinus of the pulmonary trunk

History of the Patient

Man 74 years old who died of chronic ulcerative colitis Electrocardiographic evidence of impaired intraventricular conduction noted four years prior to death but no symptoms or signs of congestive cardiac failure until a week before death Blood pressure was 150 systolic and 80 diastolic There were no cardiac murmurs (Case reported by Jordan R A *et al* *Proc Staff Meet Mayo Clin* 25 673 1950 with permission)

At necropsy the heart weighed 550 gm (normal 350 gm) Left ventricle was hypertrophied and dilated The thin walled right coronary artery arose from the right sinus of the pulmonary trunk and it terminated in the right ventricular wall The left coronary artery arose from its usual site and branched into the usual anterior descending and circumflex branches The circumflex branch followed the usual course in the left atrioventricular sulcus and then terminated as a fairly large posterior descending branch There was marked atheromatous narrowing in the anterior descending artery as well as in the circumflex artery There was a small area of fibrosis in the anterior wall of the left ventricle related to the distribution of the atheromatous anterior descending coronary artery

Principal Clinical Features of This Anomaly

Symptoms and signs of cardiac disability are usually absent when the right coronary artery arises anomalously This is in contrast to the usual case of anomalous origin of the left coronary artery





Fig. 247.—From the patient whose history is described on the preceding page. *a*, Pulmonary trunk opened through its anterior wall. The ostium of the right coronary artery (point of arrow) is shown where it lies in an aneurysm of the right pulmonary sinus. *b*, Aorta opened through its left lateral wall. The ostium of the left coronary artery may be seen at left where it has been opened (point of arrow).





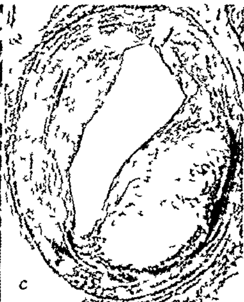


Fig. 248.—Photomicrographs of sections stained with Verhoeff elastic tissue stain counterstained with van Gieson's connective tissue stain. From the heart illustrated in Figure 4. *a*, Right coronary artery; the media is thin but resembles that of an artery; nonatheromatous fibrous intimal thickening (x15). *b*, Higher power magnification of right coronary artery showing the structure of an artery. Superficial to the well defined internal elastic lamina the intima shows fibrous nonatheromatous thickening (x3.5). *c*, Anterior descending coronary artery showing severe atheromatous change (x15).

Variations in Origin of Coronary Arteries from Aorta

Variations in manner of origin of coronary arterial supply from the aorta usually cause no functional disturbance. Occlusion of a single coronary artery can, however, give rise to very extensive myocardial infarction or sudden death.

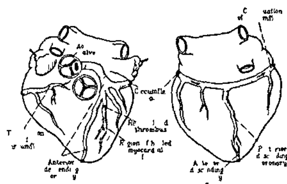


Fig. 249—Single coronary artery arising from left aortic sinus and encircling the heart. In a man 77 years old. (From Stapley, L. A., and Edwards, J. E.: *Arch. Path.* 52:470, 1951, with permission.)

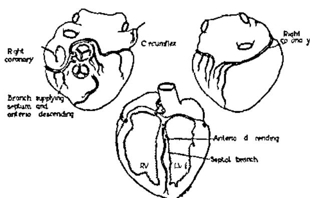


Fig. 250—Single coronary artery arising from right aortic sinus and shortly thereafter giving rise to the right coronary and left circumflex coronary arteries. Anterior descending artery arises from right coronary artery and gives off a septal branch. From a 39 year old man who died of a glioma.

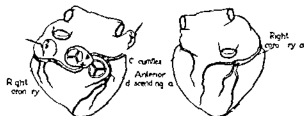


Fig. 251—Origin of both coronary arteries from left aortic sinus. In a man 77 years old.

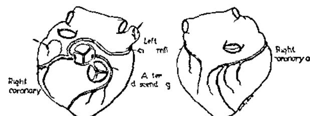


Fig. 252—Origin of left circumflex coronary artery from right coronary artery. In a man 81 years old. (Figures 250, 251 and 252 from White, N. K., and Edwards, J. E.: *Arch. Path.* 45:766, 1948, with permission.)

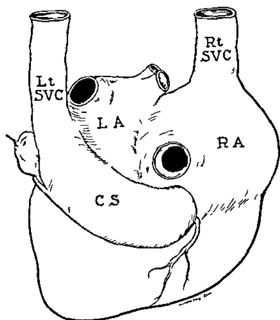
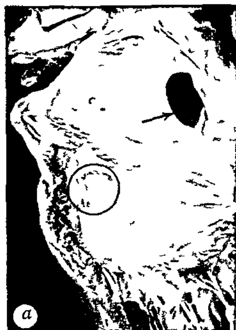


Fig 253—Diagrammatic representation of heart from behind Persistent left superior vena cava (Lt S V C) joins the left extremity of the dilated coronary sinus (C S) and the blood is carried into the right atrium (R A) L A = left atrium Rt S V C = right superior vena cava



Fig 254—Anteroposterior roentgenogram taken during cardiac catheterization The catheter has passed from the right atrium into the coronary sinus and upward into a persistent left superior vena cava (From Burchell H B *J Iowa M Soc* 38 364 1948 with permission)



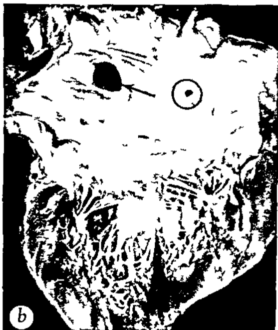


Fig 255—The heart from a man 38 years old who died of a brain tumor *a* The right side of the heart In addition to an atrial septal defect (point of arrow) there is atresia of the right atrial ostium of the coronary sinus (within circle) *b* Left side of heart Behind the atrial septal defect (point of arrow) is the opening of a vein (circle) which on one hand drains into the left atrium and on the other connects with the coronary sinus This is the major channel by which blood of a coronary sinus could enter the interior of the heart A persistent left superior vena cava is commonly associated with the atresia of the right atrial ostium of the coronary sinus but none was present in this case

Origin of an Anomalous Coronary Vessel from the Right Ventricle

FEMALE aged 4 days Intense cyanosis No murmurs or thrills Necropsy revealed pulmonary valvular atresia with intact ventricular septum An anomalous coronary vessel emerged from the right ventricle and communicated with branches of the coronary arteries (From Williams R R *et al* *Arch Path* 52 480 1951 with permission)

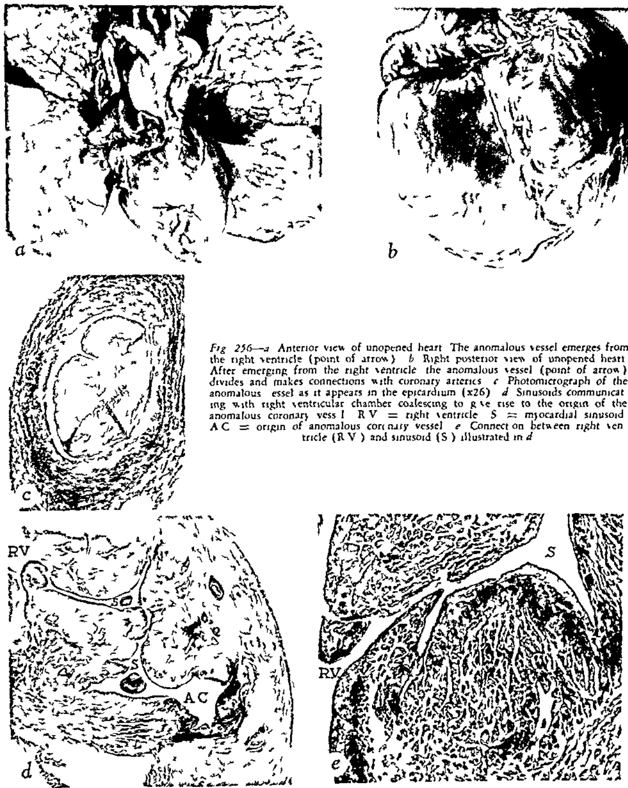


Fig 256—*a* Anterior view of unopened heart The anomalous vessel emerges from the right ventricle (point of arrow) *b* Right posterior view of unopened heart After emerging from the right ventricle the anomalous vessel (point of arrow) divides and makes connections with coronary arteries *c* Photomicrograph of the anomalous vessel as it appears in the epicardium (x26) *d* Sinusoids communicating with right ventricular chamber coalescing to give rise to the origin of the anomalous coronary vessel R V = right ventricle S = myocardial sinusoid A C = origin of anomalous coronary vessel *e* Connection between right ventricle (R V) and sinusoid (S) illustrated in *d*

Biographic Sketches



WILLIAM HUNTER

1718–1783

In a report of three cases (*Med Observations and Enquiries* 1784) William Hunter described pulmonary atresia and pulmonary stenosis associated with ventricular septal defect His emphasis on the inadequacy of pulmonary blood flow in determination of the extent of the disability is modern His clear description of the attacks of unconsciousness associated with dyspnea and cyanosis may be called a classic



THOMAS BEVILL PEACOCK

1812–1882

Among the various small yet very comprehensive monographs published near the middle of the nineteenth century that of Peacock is probably best known The clinical and pathologic correlations and the very clear illustrations are probably the best and most nearly accurate up to that time In particular the illustrations of the anomaly now called the tetralogy of Fallot are beautifully presented



WILLIAM HARVEY

1578–1657

On page 46 of *Exercitatio Anatomica de Motu Cordis et Sanguinis in Animalibus* 1628 appears such a clear description of the fetal circulation and the flow of blood through the foramen ovale that a student of congenital heart disease should have this classic work as required reading So in embryo forms whilst the lungs are idle and have no action or motion (as it there were none at all) nature makes use of both ventricles of the heart as if one for transmission of blood



JEAN BAPTISTE DE SÉNAC

1693–1770

In de Senac s detailed clinicopathologic studies of diseases of the heart *Traite de la Structure du Coeur de Son Action et de Ses Maladies* there appears a thorough discussion of the fetal circulation and a short discussion of the congenital anatomic defects of the heart De Senac has been credited (Abbott) with having first associated la maladie bleue with absence of the ventricular septum



FRANKLIN PAINE MALL

1862–1917

Franklin Paine Mall professor of anatomy in the Johns Hopkins University Medical School of Baltimore and director of the Department of Embryology of the Carnegie Institution of Washington studied structures not simply as morphologic entities but as functioning units Teacher of teachers he organized a research institute of embryology He played a prominent part in the development of many scientific publications Through the media of these publications valuable information concerning the anatomic nature of cardiac and vascular malformations and a better understanding of the embryologic basis for many of these have been supplied to the profession

JAMES HOPE

1801–1841



That James Hope was the first clinically to diagnose pulmonary stenosis with a venous arterial shunt and to predict the anatomic defects is generally accepted He closely followed a clinical case described what he thought would be found post mortem and was correct (1830) To Julius Friedrich Cohnheim (1839 1884) in 1877 goes the credit for his contribution on paradoxic embolism through a patent foramen ovale



HENRI LOUIS ROGER

1809–1891

Roger demonstrated the presence of a ventricular septal defect but without stenosis of pulmonary arteries in a boy who had not been cyanotic during life Clinical signs in association with an uncomplicated septal defect are so well described that the term maladie de Roger came into common usage He recognized the thrill and described the murmur (bruit de Roger) characteristic of a defect in the ventricular septum

VICTOR EISENMENGER

1864–1932



Eisenmenger s article appeared in 1897 under the title of *Die angeborenen Defecte der Kammerscheideuand des Herzens* In this article he described a combination of congential anomalies in which the arrangement is similar to that seen in the tetralogy of Fallot except for the fact that the pulmonary artery rather than being narrowed is either normal or dilated



ETIENNE LOUIS ARTHUR FALLOT

1850–1911

In 1888 Fallot wrote Until now clinicians have considered the diagnosis of anatomic lesions of morbus caeruleus of almost unsurmountable difficulty Although the combination of lesions known as the tetralogy of Fallot had been recognized as an anatomic entity more than 100 years before it took the next 50 years for the medical profession to appreciate the fact that this meticulous physician had paved the way for the accurate selection of cyanotic patients who might years later be benefited through surgical intervention

EDUARD SANDIFORT

1742–1814



Eduard Sandifort professor of surgery anatomy and medicine at the University of Leyden in the late eighteenth century is known as the father of pathologic iconography He has given a very clear description of the cardiac defects now known as the tetralogy of Fallot His classic description of how the finger placed in the right ventricle readily appeared in the aorta should be employed as a standard demonstration in pathologic anatomy Niels Stensen s anatomic description however antedated that of Sandifort by about 100 years



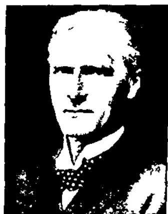
KARL ROKITANSKY

1804–1878

In Rokitansky s study *Die Defekte der Scheidewande des Herzens* 1875 the anomalies of the great vessels the defects of the atrial and ventricular septa and the theories relative to the etiology of transposition are described The illustrations of the various lesions are detailed and beautifully executed and the engraving of the anatomic defects in what we call tetralogy of Fallot is especially excellent In his study of the great vessels published in 1852 the cases of patent ductus arteriosus have an excellent clinicopathologic correlation the clinical findings often being those of his contemporary and friend Skoda

SIR ARTHUR KEITH

1866–



For many years Sir Arthur Keith pursued his studies on congenital cardiac disease mainly from the embryologic viewpoint His contributions to knowledge of the embryologic defects of the heart largely concern his observations on the normal and abnormal differentiation of the bulbus cordis However his researches were wide encompassing anatomic and physiologic correlations His descriptions of large bronchial vessels in a case of severe pulmonary stenosis are of special clinical interest



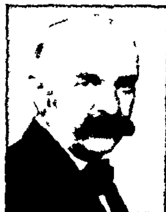
G A GIBSON

1854–1913

In Britain the characteristic murmur of a patent ductus arteriosus is still known as Gibson s murmur His repeated emphasis on the clinical diagnosis through recognition of the characteristic murmur as well as the physiologic disturbance created by it was a major contribution His article entitled *Persistence of the Arterial Duct and Its Diagnosis* published in 1900 is a classic Shortly thereafter (1907) an American surgeon John C Munro (1858 1910) prophetically referred to the possibility of ligation of a patent ductus arteriosus provided a diagnosis could be made beforehand

JULIUS TANDLER

1869–1936



Julius Tandler was the author of many works on gross anatomy as a pure science and on anatomy as applicable to the surgeon By close association with clinicians anatomy was for him a science of functioning organs To the field of congenital cardiac disease his greatest contributions were the results of his investigations on cardiac embryology His chapter on the development of the heart in Keibel and Mall s text on human embryology published in the early part of this century is still a valuable and authoritative reference



JOHN BAPTIST MORGAGNI

1682–1771

In both old and recent reviews the description of the cyanotic girl of 16 years in whom Morgagni found a pulmonary stenosis is considered to be the first good clinicopathologic study of cyanotic congenital cardiac disease It is of unusual interest because the patient had a relatively rare lesion namely pulmonary stenosis without a ventricular septal defect In spite of the rarity of the lesion it was the first type of congenital cyanotic lesion correctly diagnosed clinically (James Hope 1830) and is well illustrated by Peacock (1866)

MAUDE E ABBOTT

1869–1940



To this century s students of congenital cardiac disease particularly on the North American continent Maude Abbott s name justly comes first to mind Stimulated through an early association with Sir William Osler she in turn stimulated more extensive interest in congenital cardiac defects and their correlation with the clinical feaures Her eminence as a pathologist was widely recognized and her Atlas will long remain an authoritative source of data for studies on congenital cardiac disease

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This Book

An Atlas of Congenital Anomalies of the Heart and Great Vessels

By

JESSE EDWARDS M D *et al*

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